Progressive subcortical gliosis
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Introduction

This article includes discussion of progressive subcortical gliosis; PSG; primary subcortical gliosis; Pick disease, type II; and Pick disease, type 2. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

The author explains the clinical presentation, etiology, differential diagnosis, and diagnostic workup of progressive subcortical gliosis, a chromosome-17-linked dementia with unique pathologic features. Microscopically, the major pathologic change is a marked fibrillary astrocytosis, particularly in the area of the short cortical association tracts at the junction of cortical lamina VI and the subcortical white matter and in the subpial cerebral cortex. Overall, the survival of patients with progressive subcortical gliosis averages about 10 years and is similar to that of other types of frontotemporal dementia.

Key points

• Progressive subcortical gliosis is a rare dementing disorder resembling Pick disease but with distinctive neuropathologic features. The clinical manifestations are those of a frontotemporal dementia and overlap with those of other frontotemporal dementias.
• Progressive subcortical gliosis has an insidious onset, generally in the fifth or sixth decade. The course is progressive, generally over 5 to 15 years, but both fulminant and protracted courses occur.
• Common initial manifestations include personality and emotional changes, lack of judgment and insight, deterioration in social behavior, delusions, paranoia, auditory and visual hallucinations, and depression.
• Microscopically, the major pathologic change is a marked fibrillary astrocytosis, particularly in the area of the short cortical association tracts at the junction of cortical lamina VI and the subcortical white matter and in the subpial cerebral cortex.
• Most cases have been sporadic, but some hereditary forms are recognized, with linkage demonstrated in 1 kindred to a region on the long arm of chromosome 17 (17q21-22).
• Overall, the survival of patients with progressive subcortical gliosis averages about 10 years and is similar to that of other types of frontotemporal dementia.

Historical note and terminology

Progressive subcortical gliosis is a rare dementing disorder resembling Pick disease but with distinctive neuropathologic features. In 1949, Neumann described 3 cases under the appellation of "Pick disease, type II" (Neumann 1949). In 1967, Neumann and Cohn reported 4 additional cases and suggested the designations "primary subcortical gliosis" and "progressive subcortical gliosis" (Neumann and Cohn 1967); although neither of these terms is ideal, the latter has become established in the literature (Brun and Gustafson 2011).

Although rare reports of familial cases were reported beginning in the 1940s, it was not until the late 1980s and 1990s that it was recognized that progressive subcortical gliosis could be transmitted as an autosomal dominant trait (Currier et al 1986; Lanska et al 1991; Lanska et al 1994). Progressive subcortical gliosis was linked to a tau mutation on the long arm of chromosome 17 (Petersen et al 1995; Goedert et al 1999) and, thus, included in the "frontotemporal dementias and Parkinsonism linked to chromosome 17" group (Foster et al 1997; Brun and Gustafson 2011).

Clinical manifestations

Presentation and course

Progressive subcortical gliosis has an insidious onset, generally in the fifth or sixth decade, although individuals have
developed the disease as early as 30 years and as late as 79 years (Lanska et al 1994; Lanska et al 1998). The distribution of age of onset is similar for familial and sporadic cases. There is no apparent gender predilection.

Common initial manifestations include personality and emotional changes, lack of judgment and insight, deterioration in social behavior, delusions, paranoia, auditory and visual hallucinations, and depression (Lanska et al 1994; Lanska et al 1998). Subsequently, affected individuals develop severe memory difficulty, speech output generally declines, and word-finding problems appear. Some patients develop verbal stereotypy, echolalia, or manifestations of the human Klüver-Bucy syndrome (Lanska et al 1994).

Several cases had clinical evidence of relatively localized dysfunction, involving visual, motor, or cognitive domains (Neumann 1949; Seitelberger 1968; Auff and Seitelberger 1980; Benson et al 1988; Benson 1989; Bergmann et al 1991); in some, the changes developed rapidly over weeks, prompting a diagnosis of cerebral infarction or hemorrhage (Neumann 1949; Auff and Seitelberger 1980). Isolated sporadic cases have also been reported with posterior cerebral dysfunction or atrophy (Benson et al 1988; Benson 1989; Renner et al 2004; Borruat 2013; Ortner and Kurz 2015), or the clinical manifestations of the Steele-Richardson-Olszewski syndrome (Will et al 1988; Armstrong 2013). Terminal manifestations included profound dementia, akinetic mutism, incontinence, severe dysphagia, and extrapyramidal signs (Lanska et al 1994).

**Prognosis and complications**

The course is progressive, generally over 5 to 15 years, but both fulminant (3 months to 2 years) and protracted (spanning decades) courses occur. The duration of illness is generally longer in familial cases than in sporadic cases. The most common causes of death are pneumonia, other respiratory infections, and pulmonary emboli.

Overall, the survival of patients with progressive subcortical gliosis is similar to that of other types of frontotemporal dementia (Chiu et al 2010).

**Clinical vignette**

In his early 40s, the patient developed personality change, progressive memory impairment, and decreased activity. He was retired from his accounting work because of forgetfulness, difficulty in making decisions, and frequent errors performing simple calculations. In his late 40s, he had poor insight and judgment, impaired memory, mild word-finding problems, and difficulty performing serial subtractions. A masked face, slightly increased appendicular and axial tone, and a fine postural tremor were present, but there were no gaze abnormalities. Complete blood count, serum chemistries, thyroid function studies, and serum rapid plasma reagin were normal. Head CT showed mild cerebral atrophy with ventriculomegaly. Radioisotope cisternography was normal. EEG showed well-modulated 10-Hz alpha activity posteriorly, but frequent, rhythmic, low-amplitude delta transients were observed in the right anterior and right midfrontal areas. Cerebrospinal fluid examination showed an opening pressure of 180 mm H2O, a protein level of 27 mg/dL, a glucose level of 55 mg/dL, and no cells.

At age 50, the patient had decreased verbal fluency, mild comprehension impairment, and word-finding difficulty with semantic paraphasic errors. Over the next several years, he became more compulsive, regimented, and perseverative. His affect became progressively blunted. He lost interest in sexual intercourse, was incontinent, and began eating excessively. He stopped speaking spontaneously but had prominent echolalia. Later he developed severe dysphagia and became bed-bound and emaciated. He died at age 58 years from aspiration.

**Biological basis**

**Etiology and pathogenesis**

Most reported cases of progressive subcortical gliosis have been sporadic, but there are several reports of familial progressive subcortical gliosis (Hassin and Levitin 1941; Neumann and Cohn 1967; Khoubesserian et al 1985; Currier et al 1986; Lanska et al 1989; Lanska et al 1991; Lanska et al 1994; Swerdlow et al 2009). In several of the initial reports of familial progressive subcortical gliosis, family history information is sketchy, and detailed clinical information and pathological confirmation are available only for the index cases (Hassin and Levitin 1941; Neumann and Cohn 1967; Khoubesserian et al 1985). In a preliminary report in 1986, Currier and colleagues first demonstrated that progressive subcortical gliosis could be transmitted as an autosomal dominant trait (Currier et al 1986). In a large kindred of 90 individuals spanning 6 generations, 21 of 65 members in 4 consecutive generations may have been
affected, and 5 members from 2 generations were proved to have been affected on the basis of autopsy (Lanska et al 1991; Lanska et al 1994). Lanska and colleagues subsequently confirmed these findings in a second kindred of 80 individuals spanning 4 generations. 10 members of 30 members in 3 consecutive generations may have been affected by the disease, and 2 siblings were proved to have been affected on the basis of autopsy (Lanska et al 1989; Lanska et al 1991; Lanska et al 1994). In 1995, Petersen and colleagues suggested that familial progressive subcortical gliosis may be a prion disease (Petersen et al 1995), but this conclusion was later retracted (Gambetti 1997).

Linkage has been demonstrated in 1 of the kindreds reported by Lanska and colleagues (Lanska et al 1994) to a region on the long arm of chromosome 17 (17q21-22) (Petersen et al 1995; Foster et al 1996; Foster et al 1997; Wilhelmens et al 1996), a site previously associated with the disinhibition-dementia-parkinsonism-amyotrophy complex (Lynch et al 1994; Wilhelmens et al 1994; Wilhelmens et al 1996). As early as 1995, it was recognized that candidate genes in the region include microtubule-associated protein-1 (known as tau) (Petersen et al 1995). Subsequently, a tau mutation at position +16 of the intron after exon 10 has been identified (Goedert et al 1999) in 1 kindred with progressive subcortical gliosis (Lanska et al 1994; Petersen et al 1995). The mutation destabilizes a predicted stem-loop structure and leads to an overrepresentation of the soluble 4-repeat tau isoforms that assemble into wide, twisted, ribbon-like filaments. Overproduction of 4-repeat tau is probably the primary effect of the progressive subcortical gliosis mutation. It may lead to an excess of 4-repeat tau over available binding sites on microtubules, resulting in the hyperphosphorylation of tau and its assembly into filaments (Krishnamurthy and Johnson 2004).

Some other conditions included in the "frontotemporal dementias and Parkinsonism linked to chromosome 17" group (Foster et al 1997), but with clinicopathologic differences from progressive subcortical gliosis (Foster et al 1997; Mann 1998; Miyamoto et al 2001), have mutations in the tau gene (Clark et al 1998; Dumanchin et al 1998; Hong et al 1998; Hutton et al 1998; Poorjak et al 1998; Spillantini et al 1998a; Spillantini et al 1998c; Morris et al 1999; Miyamoto et al 2001; Tsuboi et al 2002; Hughes et al 2003; Soliveri et al 2003; van Herpen et al 2003; van Swieten et al 2004; Wilhelmens et al 2004; Boeve et al 2005; Bronner et al 2005; Zarranz et al 2005; Doran et al 2007; Kowalska 2009; Kimura et al 2016). Several studies have identified various missense mutations, silent mutations, and amino acid deletions in the tau gene in patients with frontotemporal dementias (Hutton et al 1998; Bugiani et al 1999; Delisle et al 1999; D’Souza et al 1999; Hasegawa et al 1999; Iijima et al 1999; Murrell et al 1999; Rizzu et al 1999; Arima et al 2000; Lippa et al 2000; Rizzini et al 2000; Spillantini et al 2000a; Spillantini et al 2000b; Stanford et al 2000; Stanford et al 2004; Lee 2001; Miyasaka et al 2001; Morris et al 2001; Neumann et al 2001; Pastor et al 2001; Hayashi et al 2002; Pickering-Brown et al 2002; Rosso et al 2002; Saito et al 2002; Tsuboi et al 2002; Soliveri et al 2003; van Herpen et al 2003; Kobayashi et al 2004; Kondo et al 2004; Ostojic et al 2004; Passant et al 2004; Boeve et al 2005; Bronner et al 2005; Zarranz et al 2005; Doran et al 2007; Niblock and Gallo 2012). Mutations have also been described at positions +3, +12, +13, +14, and +16 of the intron after exon 10 of the tau gene, and at the intron 10+11-splice site (Hutton et al 1998; Spillantini et al 1998b; Conneally et al 2000; Yasuda et al 2000; Lee et al 2001; Miyamoto et al 2001; Doran et al 2007; Kowalska 2009). Detailed tau pathology has been reported for the kindred with the +3 mutation, also known as "familial multiple system tauopathy with presenile dementia" (Spillantini et al 1997; Spillantini et al 1998b); this kindred has neuronal and glial tau pathology similar to that of progressive subcortical gliosis. It is also characterized by the presence of wide, twisted ribbons made of 4-repeat tau isoforms and an overproduction of soluble 4-repeat tau, indicating that the different mutations in the intron after exon 10 cause disease through the same mechanism. These findings firmly establish that the overproduction of 4-repeat tau alone is sufficient to lead to filamentous tau pathology and a dementing disease (Spillantini et al 2000a).

How changes in the ratio of 3-repeat tau to 4-repeat tau produce neuronal and glial dysfunction and cell death is unknown. 3-repeat tau and 4-repeat tau may bind to distinct microtubule sites, and an altered ratio of these isoforms may disrupt microtubule function (Spillantini et al 2000a; Lee et al 2001). Also, overproduction of 4-repeat tau may simply produce an excess of free cytosolic tau, which then becomes hyperphosphorylated and is assembled into pathologic filaments (Lee et al 2001). Other mutations alter the binding of tau to microtubules and interfere with its ability to promote microtubule assembly (Spillantini et al 2000a; Lee et al 2001).

The existence of tau mutations with distinct pathogenetic mechanisms may explain the phenotypic heterogeneity of atypical dementias that previously led to their classification into separate disease entities.

A separate kindred with autosomal dominant subcortical gliosis presenting as a frontotemporal dementia, but without
tau- or ubiquitin-containing inclusions and without ubiquitin or TDP-43 staining, has been reported by Swerdlow and colleagues (Swerdlow et al 2009). Symptom onset was generally in the fifth or sixth decade, but some individuals did not become symptomatic until their eighth decade.

The brains of progressive subcortical gliosis cases are moderately to severely atrophic, usually with preferential involvement of the white matter of the frontal and temporal lobes (Lanska et al 1994), but several cases affected mainly the frontal lobes, and 1 sporadic case affected predominantly the parietal and occipital lobes (Benson et al 1988; Benson 1989). Most often the atrophy is symmetric (Lanska et al 1994), but in several cases, 1 side was more affected than the other (Neumann 1949; Yano et al 1960; Seitelberger 1968; Verity and Wechsler 1987).

Microscopically, the major pathologic change is a marked fibrillary astrocytosis, particularly in the area of the short cortical association tracts at the junction of cortical lamina VI and the subcortical white matter and in the subpial cerebral cortex (Neumann and Cohn 1967; Seitelberger 1968; Masse et al 1981; Seitelberger et al 1983; Verity and Wechsler 1987; Lanska et al 1994). The gliosis is most pronounced in the frontal and temporal lobes, occurs in the absence of identifiable myelin loss, and is out of proportion to the mild neuronal loss in the cortex (Verity and Wechsler 1987; Lanska et al 1994). Lesser degrees of gliosis are observed in the deep white matter, basal ganglia, thalamus, brainstem tegmentum, cerebellum, and central gray matter of the upper cervical spinal cord (Lanska et al 1994). In addition, variable protoplasmic astrocytic proliferation is evident in all layers of the cerebral cortex (Seitelberger 1968; Masse et al 1981; Seitelberger et al 1983; Bergmann et al 1991; Lanska et al 1994).

Laminar spongiosis has been variably reported in laminae II and III of the cerebral cortex in cases of progressive subcortical gliosis (Neumann and Cohn 1967; Seitelberger 1968; Masse et al 1981; Seitelberger et al 1983; Verity and Wechsler 1987; Lanska et al 1994). The spongiosis may be focal and probably occurs late in the disease. It generally spares the deeper layers of the cortex, as well as the basal ganglia, thalamus, brainstem, and cerebellum (Lanska et al 1994). Ultrastructural studies have shown areas of clarification and frequent vacuoles in large postsynaptic dendrites surrounded by glycogen and mitochondria (Masse et al 1981). This spongiosis has been attributed variously to neuronal loss, loss of dendritic arborizations of neurons that expand in the second layer, and pathologic alteration and disintegration of astrocytes (Masse et al 1981; Seitelberger et al 1983; Verity and Wechsler 1987). The spongiosis of progressive subcortical gliosis is similar to that in other neurodegenerative diseases (eg, Pick disease, Alzheimer disease), but it differs from the spongiform change of Creutzfeldt-Jakob disease, which is pancortical, most prominent in the deeper layers, and frequently involves the thalamus, basal ganglia, brainstem, and cerebellum (Masters and Richardson 1978; Lanska et al 1994).

Neuronal cytoskeletal inclusions (Pick bodies, neurofibrillary tangles, Lewy bodies) are absent in progressive subcortical gliosis (Lanska et al 1994). Several cases had occasional balloon neurons in the cerebral cortex (Neumann 1949; Yano et al 1960); however, most authors did not identify such cells. Although considered 1 of the histopathologic hallmarks of Pick disease, balloon neurons are nonspecific and do occur in several other neurodegenerative disorders.

In several cases, the pars compacta of the substantia nigra exhibited moderate to marked degenerative changes (Lanska et al 1994). There was no apparent neuronal loss or gliosis in the nucleus basalis of Meynert in the cases wherein this was examined (Lanska et al 1994).

Studies in a transgenic mouse model of frontotemporal dementia and parkinsonism linked to chromosome 17 indicate that the L-dopa-resistant parkinsonism in this tauopathy results from early loss of dopaminoreceptive neurons in the striatum (Chiba et al 2012). Accumulation of mutant tau leads to neurofibril degeneration and apoptosis through caspase-3 activation (Chiba et al 2012). In the late stages, dopaminergic neurons and dopaminoreceptive neurons decreased equally (Chiba et al 2012).

Alterations in pronerve growth factor and brain-derived neurotrophic factor have been identified in non-Alzheimer disease tauopathies (Belrose et al 2014). Neurotrophin dysregulation as a result of tau pathology may represent a mechanism by which tau produces neurotoxicity in progressive subcortical gliosis and other non-Alzheimer dementias (Belrose et al 2014).
Even though the hippocampus is not the primary pathologic site in progressive subcortical gliosis and other tauopathies, there are nevertheless disease-specific regional selective neuronal vulnerabilities that can be demonstrated in the hippocampus for these disorders (Milenkovic et al 2014).

Epidemiology"

The frequency of progressive subcortical gliosis in the general population is unknown. It is thought to be a rare disorder, but limited data from autopsy series and brain banks suggest that progressive subcortical gliosis is more common than generally recognized. In autopsy series of demented individuals, progressive subcortical gliosis has been identified in as many as 4% to 7% of cases (Sulkava et al 1983; Boller et al 1989; Gearing et al 1995). Cases with the disorder are present in most established brain banks; in general, the proportion of cases with progressive subcortical gliosis in established brain banks is similar to the proportions with progressive supranuclear palsy, Pick disease, or Creutzfeldt-Jakob disease. Nevertheless, the relative frequency of progressive subcortical gliosis may be overestimated by such data because cases with early onset and unusual features are perhaps more likely to be autopsied.

Differential diagnosis

The clinical manifestations of progressive subcortical gliosis are those of a frontotemporal dementia and overlap with those of Pick disease and other frontotemporal dementias, including Alzheimer disease, corticobasal ganglionic degeneration, Binswanger disease, and similar conditions (Kim et al 1995; Wada et al 2006; Bugiani 2007; Tacik et al 2015).

Although in many cases the clinical manifestations of progressive subcortical gliosis are similar to those of Pick disease and Alzheimer disease, progressive subcortical gliosis can be distinguished on pathologic grounds (Neumann 1949; Neumann and Cohn 1967; Lanska et al 1994; Lanska et al 1998; Kim et al 1995). Progressive subcortical gliosis shares with Pick disease and other frontotemporal dementias a predilection for the frontal and temporal poles, whereas in Alzheimer disease, the posterior temporal and parietal lobes are preferentially involved. The atrophic process in progressive subcortical gliosis is not sharply demarcated, whereas in Pick disease, there are typically circumscribed areas of severe atrophy with an abrupt transition between severely affected and less affected areas.

In progressive subcortical gliosis, cortical neuron loss is mild or inconspicuous, and a characteristically distributed astrogliosis is most prominent subcortically and in the deep cortical laminae. In both Pick disease and Alzheimer disease, cerebral cortical neuron loss is prominent and is associated with a commensurate degree of cortical gliosis and little subcortical gliosis. Cerebrocortical neuronal inclusions are uniformly absent in progressive subcortical gliosis but are pathologic hallmarks of Alzheimer disease and Pick disease, seen even in early or incipient cases (Miki et al 2014).

Despite these distinctions, the differentiation of progressive subcortical gliosis from Pick disease may be difficult, and it is likely that some authors lump progressive subcortical gliosis with putative subtypes of Pick disease without Pick bodies (eg, Pick disease "type C" of Constantinidis and colleagues) (Constantinidis et al 1974; Tissot et al 1985; Ikeda 2000).

Compared with other forms of frontotemporal dementia (Hodges et al 2004; Kertesz et al 2005), progressive subcortical gliosis appears to have a distinctive clinicopathologic profile (Lanska et al 1994; Foster et al 1997; Mann 1998), but several specific forms of frontotemporal dementia, including progressive subcortical gliosis, have now been linked to chromosome 17 (Foster et al 1997; Conneally et al 2000).

Thalamic degenerations involve similar neural structures and have histopathologic similarities to progressive subcortical gliosis (Oda 1976; Bergmann et al 1991; Lanska et al 1994). Primary thalamic degenerations are frequently accompanied by secondary gliosis in the cerebral cortex, subcortical white matter, basal ganglia, and various brainstem nuclei. Thalamic gliosis and neuron loss occur in progressive subcortical gliosis (Yano et al 1960; Neumann and Cohn 1967; Bergmann et al 1991); however, they apparently develop independently of the other pathologic changes, because: (1) there is prominent gliosis of the short ipsilateral cortical association fibers that have no direct thalamic connections; (2) there is neither demyelination nor axon loss in the thalamic radiations; (3) gliosis in the cerebral cortex generally spares laminae III and IV, where the majority of thalamocortical projection fibers terminate (Lanska et al 1994).

Some of the "atypical" dementias have features in common with progressive subcortical gliosis. Although progressive
subcortical gliosis and "frontal lobe degeneration of non-Alzheimer type" are clinically similar (Gustafson 1987), pathologic differences exist between the 2 conditions (Brun 1987; Englund and Brun 1987; Lanska et al 1994). In frontal lobe degeneration, little atrophy occurs, and brain weights are generally greater in frontal lobe degeneration than in progressive subcortical gliosis. In addition, the pathologic changes of frontal lobe degeneration are more evident in the cerebral cortex rather than in the subcortical areas, with prominent cortical neuron loss and mild gliosis involving mainly the outer cortex. Finally, areas typically involved in progressive subcortical gliosis (basal ganglia, thalamus, substantia nigra, inferior olivary nucleus) are either not affected or are much less affected in frontal lobe degeneration. Several other reported cases of familial (Kim et al 1981; Moossy et al 1987; Knapman et al 1990) and sporadic (Torack and Morris 1986; Knapman et al 1990) atypical non-Alzheimer dementia had clinical and pathologic similarities to progressive subcortical gliosis, but in all of these cases there was prominent cortical neuron loss in addition to variable degrees of gliosis.

**Diagnostic workup**

Cerebral imaging is normal early in the course but later shows generalized cerebral atrophy, sometimes preferentially affecting the frontal and opercular areas. Electroencephalograms remain normal except for mild slowing late in the illness. Screening for tau gene mutations in patients with frontotemporal dementia is still largely a research tool and is unlikely to identify pathogenic mutations in sporadic cases (Stanford et al 2004).

**Management**

Management is supportive. Although antipsychotic medications have been used to control hallucinations and paranoia, such agents may increase the extrapyramidal dysfunction that is commonly present in the later stages of the disorder.

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**References especially recommended by the author or editor for general reading.

ICD and OMIM codes

ICD codes

ICD-9:
Specific senile psychotic conditions: 290.8
Pick disease: 331.11

ICD-10:
Unspecified dementia, including senile dementia: F03
Dementia in Pick disease: F02.0
Pick disease: G31.0

OMIM numbers

Pick disease, type II: #172700
Familial progressive subcortical gliosis: #221820

Profile

Age range of presentation

19-44 years
45-64 years
65+ years

Sex preponderance

male=female

Family history

family history may be obtained

Heredity

heredity may be a factor
heredity typical
autosomal dominant

Population groups selectively affected

none selectively affected

Occupation groups selectively affected

none selectively affected

Differential diagnosis list

Pick disease
Alzheimer disease
other frontotemporal dementias
corticobasal ganglionic degeneration
Binswanger disease
thalamic degenerations
“atypical” dementias
frontal lobe degeneration of non-Alzheimer type

**Associated disorders**

Atypical dementia
Disinhibition-dementia-parkinsonism-amyotrophy complex
Frontal lobe dementias
Frontotemporal dementia
Frontotemporal dementia with parkinsonism linked to chromosome 17
Nonspecific dementia
Steele-Richardson-Olszewski syndrome
Tauopathy
Thalamic dementia

**Other topics to consider**

Frontotemporal dementia
Tauopathies with dementia and parkinsonism

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