Hemorrhagic Necrotic Schwannoma Presenting as Purulent Meningitis

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ABSTRACT: ***Background:*** Intracranial schwannoma involving the XIIth cranial nerve is rare. We report an unusual clinical presentation and pathological verification of a schwannoma, which had become haemorrhagic and necrotic, simulating acute purulent meningitis. ***Methods:*** A literature review of intracranial tumors presenting as acute purulent meningitis, with emphasis on schwannomas, was undertaken. ***Results:*** Few cases of hypoglossal schwannoma have been reported; the association with purulent meningitis has not been previously documented. ***Conclusion:*** In patients presenting with sterile purulent meningitis, a necrotic tumor should be considered as a possible etiology.

RÉSUMÉ: Schwannome nécrotique et hémorragique dont la présentation est celle d'une méningite purulente. ***Introduction:*** Le schwannome intracrânien impliquant le douzième nerf crânien est rare. Nous rapportons un cas de schwannome avec vérification anatomopathologique dont la présentation clinique était inhabituelle et qui était devenu hémorragique et nécrotique, simulant une méningite purulente aiguë. ***Méthodes:*** Nous avons effectué une revue de la littérature sur les tumeurs intracrâniennes dont le mode de présentation était celui d'une méningite purulente aiguë en mettent l'emphase sur le schwannome. ***Résultats:*** Peu de cas de schwannome de l'hypoglosse ont été rapportés et l'association avec une méningite purulente n'a pas été documentée antérieurement. ***Conclusion:*** Chez les patients qui ont un tableau clinique de méningite purulente aseptique, une tumeur nécrotique devrait être considérée comme une étiologie possible.

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Schwannomas account for 8%[1,2] of intracranial tumors. While their clinical presentation is identical to other intracranial tumors, such as features of raised intracranial pressure or subacute neurological symptoms, acute meningeal symptoms can also herald the presence of a neoplasm. This paper reviews this unusual presentation secondary to an intracranial schwannoma in a patient without any stigmata of neurofibromatosis.

Case Report

A 32-year-old male, smoker and asthmatic (not on steroids) presented with a 5 day history of sudden onset of sub-occipital headache with nausea, vomiting, fever, chills and neck stiffness. He also complained of dysphagia, hoarseness and noted that his right pupil was "bigger". On examination, he was a mildly obese individual, looking unwell, with a low grade temperature (38.7°C) and stable vital signs. He was alert, following commands and had moderate nuchal rigidity and mild right Horner's syndrome with both pupils reactive to light. His voice was hoarse. The fundoscopic examination was normal. He had no evidence of long tract signs, or cutaneous stigmata of neurofibromatosis.

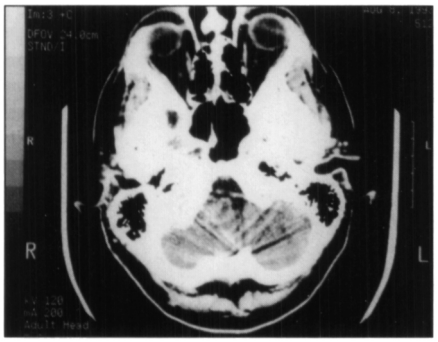

Figure 1: *Axial contrast enhanced CT scan showing ring-enhancing lesion adjacent to lower brainstem.*

The initial investigations were unrevealing. Prior to a lumbar puncture (LP), a non-enhanced CT scan of the head was obtained which was interpreted as normal. Based on cerebrospinal fluid (CSF) findings (Table 1), intravenous ceftriaxone, ampicillin, and flagyl were empirically initiated. Decadron was started for secondary focal neurological deficit with brainstem findings (cranial nerve abnormalities).

Course in Hospital

The patient became afebrile with subjective improvement but continued to complain of dysphagia. Following a 10 day course of antibiotics the LP was repeated and was not indicative of any interval change, while the repeat CT scan with IV contrast done on day two showed a ring-enhancing lesion adjacent to the lower brainstem (Figure 1). The anatomical location was verified by an MRI with intravenous gadolinium (Figure 2). Subsequent neurological examination revealed a mild right hypoglossal nerve palsy with early atrophy and fasciculations of the tongue. There was also diminished pin-prick perception over the left hemi-body, right Horner's syndrome and right vocal cord paresis. A presumptive diagnosis of a brain abscess was made. The differential diagnosis at this juncture was of an extra-axial empyema, hemorrhagic tumor, or a partially thrombosed aneurysm. Cerebral angiography excluded the latter.

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Due to the persistence of symptoms, a diagnostic procedure was performed.

Operative Findings

A sub-occipital craniectomy was performed and upon retracting the right cerebellar hemisphere, an oval, brownish tumor, arising from the right hypoglossal nerve and compressing the medulla, was noted. The lower cranial nerves were displaced laterally (Figure 3). A gross total resection of the tumor was undertaken. Postoperatively, the patient made a rapid, uneventful recovery and was discharged within a week of surgery.

Table 1:

Peripheral blood		Cerebrospinal fluid	
At admission			
white blood cells	17.7 x10E9/L	white blood cells	2290 x10E6/L
lymphocytes	2.0 x10E9/L	red blood cells	700 x10E6/L
monocytes	1.1 x10E9/L	Glucose	3.2 MMOL/L
neutrophils	14.6 x10E9/L	Protein	1761 MG/L
Glucose	6.2 MMOL/L	Acid fast bacilli	nil
		Fungi	nil
		Pseudomonas fluorescens	present
Pre-operative			
white blood cells	16.5 x10E9/L	white blood cells	1820 x10E6/L
		red blood cells	5 x10E6/L
		neutrophils	1347 x10E6/L
		lymphocytes	382 x10E6/L
2 weeks Post-operative			
		white blood cells	44 x10E6/L
		red blood cells	6 x10E6/L
		neutrophils	0 x10E6/L
		lymphocytes	38 x10E6/L
		monocytes	6 x10E6/L
		Glucose	2.0 MMOL/L
		Protein	1784 MG/L
		No growth after 3 days	

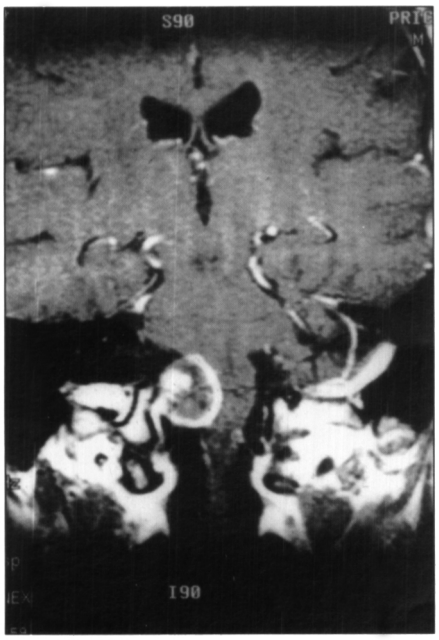

Figure 2: *Coronal MR scan with Gadolinium showing ring-enhancment of cystic lesion compressing brainstem.*

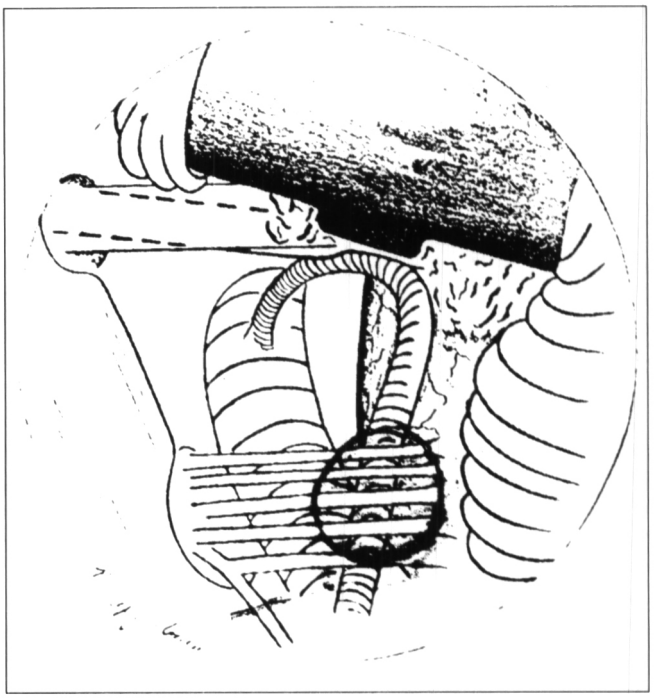

Figure 3: *Diagramatic representation of tumor when exposed.*

Pathology

Grossly, the tumor was necrotic with old hemorrhagic regions surrounded by xanthochromic and gliotic areas. Culture specimens taken from within the tumor remained sterile and frozen pathological sections obtained at surgery confirmed the presence of a tumor. Permanent microscopic sections showed schwannoma with extensive tumor necrosis. There were numerous hyalinized blood vessels, some of them thrombosed. In some necrotic areas, the cytoarchitecture was maintained and nuclear palisading could still be discerned (Figure 4). Fresh and old hemorrhages (suggested by iron pigment deposition) were apparent.

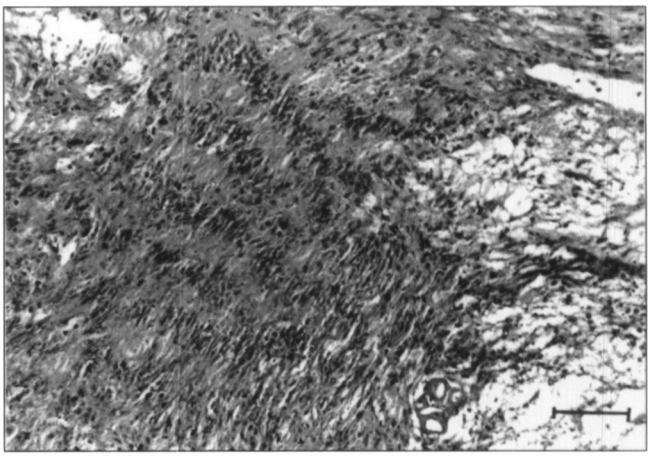

Figure 4: *Tumor with spindle shaped cells forming nuclear palisaides. There is early necrosis with nuclear fragmentation (Haemotoxylin - eosin; Bar = 64 um).*

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Discussion

This patient demonstrated 3 unusual clinical features, namely: 1) a purulent meningitic picture; 2) hypoglossal schwannoma; 3) necrotic, hemorrhagic schwannoma.

The term "aseptic meningitis" was introduced by Ayer[3] in 1920, to describe the meningeal reaction which followed aseptic irritation exhibited by the presence of lymphocytes, macrophages, plasma cells and fibroblasts in the subarachnoid space. This term was adopted by Walgren[4] in a publication in 1925. Other terms have been used since then including Decker and Gross's[5] description of a meningeal reaction secondary to a ruptured dermoid cyst, which they referred to as "chemical meningitis".

These synonyms both indicate meningeal responses resulting from leakage of neoplastic contents into the CSF pathway from cranial or spinal cystic tumors. It would appear that implicit in these terms is a predominant mononuclear pleocytosis, which is the usual CSF cellular response frequently observed in aseptic meningitis. An elevated polymorphonuclear (PMN) cell count is an unusual response, but this was first documented by Brown[6] and later by Bilger.[7] Soffer[8] reported 3 cases of primary brain tumors simulating purulent meningitis. The first two cases harboured cerebellar and fourth ventricular ependymoma and the third patient a temporal lobe astrocytoma. All those patients presented with clinical and laboratory findings consistent with purulent meningitis. Diagnosis was made by surgery in the first two cases and at autopsy in the last case. Except for an isolated instance of a CSF smear showing gram positive cocci, which was not upheld by subsequent daily smears or cultures,[6] the CSF cultures remained sterile in all those cases. A similar finding occurred in our case, where an isolated culture of "Pseudomonas fluorescens", a contaminant, misled and delayed the appropriate treatment of the patient.

Numerous reports of brain tumors presenting as a chemical meningitis have been documented. Lunardi[9] reviewed 35 cases previously reported in the literature, in addition to his own case report, and noted that embryonal tumors and malignant gliomas with cystic components were the principal diagnosis in the series. In fact, 50% of the tumors were dermoid/epidermoid, occurring predominantly in children 8 years old and under. There was no case of schwannoma in the series.

Nerve sheath tumors comprise less than 8% of intracranial neoplasms, more commonly involving cranial nerves VIII, VII and V.[1,2] There is a predominance in the female gender; Berger et al.[10] reported a ratio of 2.6:1. Cranial nerve XII schwannomas are very uncommon.[10-25] Our case is also remarkable because, to the best of our knowledge, this is the only documented case of intracranial schwannoma manifesting with a clinical picture of purulent meningitis. A review of these cases shows that while long standing signs of cranial nerve XII palsy were present in all except one,[10] this was not the presenting complaint. The most common complaint was headache and meningismus, with cranial nerve palsy and brain stem compression being less frequent.

Asari[26] reviewed 25 patients with intraspinal and intracranial hemorrhagic schwannomas ranging in size from 2.8 cm to 3.8 cm. Cranial nerve VIII was involved in 80% of the cases, while there was no instance of cranial nerve XII involvement. He classified the pattern of hemorrhage as intratumoral, subarachnoid or combined. Intratumoral hemorrhage occurred in half the cases and combined in a third. Our case showed a combined pattern. The mechanism of hemorrhage remains obscure, in contrast to malignant tumors where intratumoral vessel occlusion with distal necrosis and erosion of vessels may increase the tendency to bleed.[26] Histological sections of schwannomas have shown multi-focal areas of sinusoidal dilatations and thick hyalinized vessel walls, presumably predisposing them to rupture. A possible association with hypertension has been considered but not found in other studies.[26,27] Size appears to be an important factor as all the tumors that bled were larger than 2.8 cm.[26]

Treatment and Outcome

In the past, operative excision of hypoglossal schwannoma was attendant with high mortality.[28] Most causes of operative morbidity and mortality were related to respiratory difficulties and severe post-operative neurological deficits, requiring prolonged ventilation and subsequent tracheotomy. An important contributing factor may be delay in diagnosis. However, standard exposure by a sub-occipital or lateral approach,[29] with adequate monitoring and the use of an operative microscope, have decreased these complications and, as with our patient, ensure a smooth and uneventful postoperative course.

Conclusion

We believe that the concept of further investigation of patients with sterile purulent meningitis should be re-emphasized. Imaging studies done prior to lumbar puncture to rule out the presence of brain abscess in purulent meningitis should not fail to include the possibility of a necrotic tumor in the differential diagnosis. Subsequent treatment may be significantly altered, as surgical intervention may be contemplated sooner.

Secondly, diagnosis of purulent meningitis in a patient with minimal alteration in level of consciousness should raise doubts as the sole explanation of the patient's illness. However, we recognize that in the early phase of meningitis, alteration in the level of consciousness may be minimal.

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