Case Report

Extracranial Meningioma Presenting as a Neck Mass in a Patient with Underlying Ankylosing Spondylitis

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ABSTRACT. Extracranial meningiomas are rare. They account for only 2% of meningiomas compared to the much more common intracranial site. We describe a rare case of extracranial meningioma presenting as a neck lump in a patient with preexisting neck disease due to ankylosing spondylitis. Extracranial meningioma should be considered in the differential diagnosis of atypical neck lumps. Investigations including magnetic resonance imaging and histopathology are discussed, as well as the results of a literature review on extracranial meningiomas. (J Rheumatol 2006;33:1883–5)

Key Indexing Terms:
MENINGIOMA NECK ANKYLOSING SPONDYLITIS

Meningioma is a well recognized tumor of the central nervous system (CNS) accounting for 20% of all intracranial tumors; it is the second most common tumor of the CNS[1,2]. Meningiomas are more common in women than in men, with a ratio of 2:1 and peak incidence at age 45 years[3]. Meningiomas have been described in children, where they tend to be more aggressive, multiple, and occasionally malignant[4]. At autopsy asymptomatic meningiomas are found in 1.4% of cases and are usually isolated findings, except when associated with Von Recklinghausen disease[2,3,5]. This same trend is also seen in primary extracranial meningioma, which also shows a strong female predominance[3].

Primary extracranial tumors are often difficult to diagnose and must be thoroughly evaluated in order to rule out an intracranial component. In the head and neck area, extracranial meningiomas have been described in various sites, such as the temporal bone, middle ear[3,6-8], nasal cavity[9], parotid gland[10], and paranasal sinus[1,6].

CASE REPORT

A 51-year-old woman with a long-standing history of ankylosing spondylitis (AS) was referred from rheumatology with a 6-month history of a swelling in the right posterior triangle. There was very limited neck movement as a result of AS, but the swelling was not associated with any pain.

On examination there was a 3 × 5 cm lump, which was hard in consistency and fixed, with no focal neurology (Figure 1). Further ENT examination was unremarkable. An incisional biopsy was performed, and histological examination revealed features consistent with meningioma (Figure 2). There were no histological features suggestive of a malignant melanoma. Overall, the morphological features were in keeping with a meningioma.

Magnetic resonance imaging (MRI) was required to differentiate between a peripheral extension of an intracranial meningioma and the less common occurrence of an extracranial meningioma. Head and neck MRI showed a mass measuring 2.2 cm in width, 3 cm in anteroposterior diameter, and 4.8 cm in vertical diameter. The mass was lying in the posterior triangle of the right neck, deep to the levator scapula muscle, which the mass lifted laterally. It was insinuating into several intravertebral foramina, but did not occlude any of these foramina (Figure 3). There was no evidence of any extension into the spinal canal or intracranial involvement.

Overall histopathology confirmed meningioma, and MRI showed that this was the much rarer extracranial type. The lesion is benign with no mitotic activity. Our patient has therefore been initially managed with a "wait and watch" policy, as she is pain-free with no neurological symptoms.

DISCUSSION

Meningiomas are slow-growing lesions that represent about 20% of all intracranial tumors; however, primary extracranial meningiomas are relatively infrequent.

Considered by most to be benign hamartomas, they arise from meningothelial arachnoid cells, which are derived from the neuroectoderm. These cells are found in clusters at the tip of arachnoid membrane through the walls of dural sinuses into the sinus lumen, where they absorb cerebrospinal fluid. They are concentrated along the major dural sinuses and their tributary veins[2,4-6,11].

Meningiomas have a variety of easily recognizable histomorphological features. The cells appear as round, polygonal, ovoid epithelial, or endothelial types of cells. The nuclei have a delicate appearance and occasionally large nuclei may be present but are usually uniform. Mitoses are extremely rare[6]. The most common histologic classification of these tumors is based on the predominant cell type and includes 5 groups: syncytial, fibroblastic, transitional, angioblastic, and sarcomatous[1,3,8].

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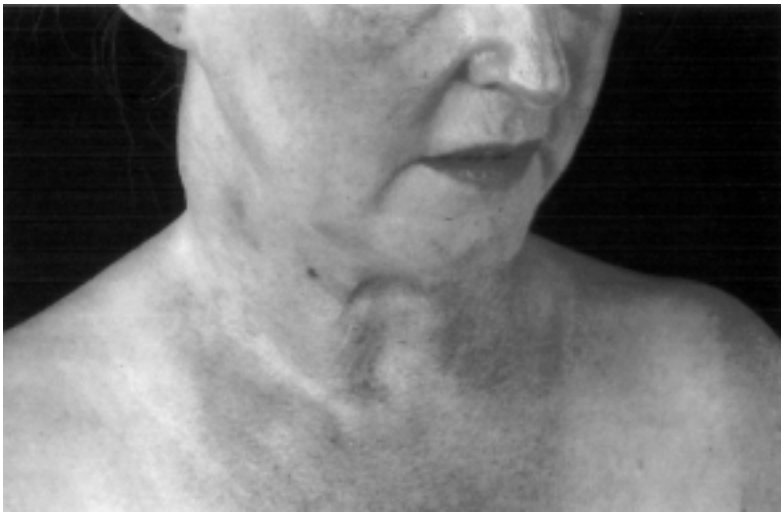

Figure 1. The right posterior triangle neck lump.

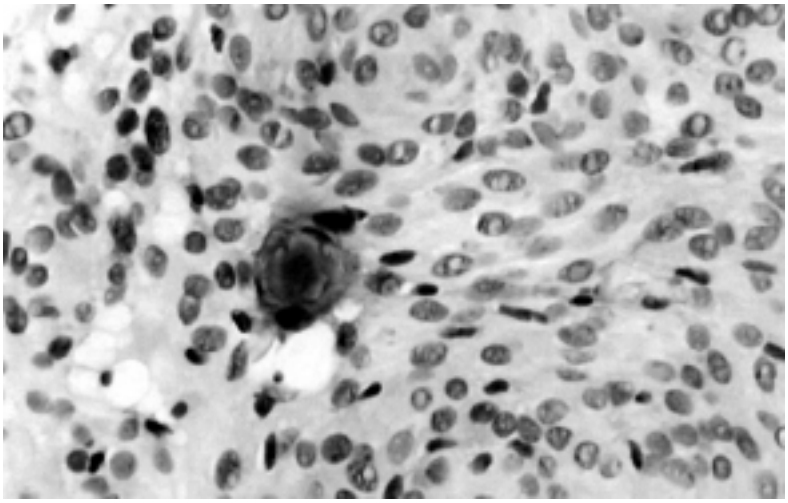

Figure 2. Histopathological specimen shows nests of epithelioid cells with a whorled pattern containing psammoma body formation. There is no evidence of cellular pleomorphism and no mitotic activity. Immunocytochemistry showed focal EMA positivity with no staining for cytokeratins or S100. Hematoxylin and eosin; original magnification ×400.

Many theories have been suggested for the mechanism of occurrence of extracranial arachnoid cells: Arachnoid cellular rests have been proposed to be formed by aberrant migration of arachnoid cells during embryogenesis[2,8,12]. Alternatively, neural crest cells, schwann cells, arachnoid cells, and fibroblasts, which are mesodermal in origin, are cell types with potential to give rise to meningiomas[2,8].

Extracranial meningiomas are categorized in 4 groups, as described by Hoye, *et al*[13]: (1) primarily intracranial tumors with extracranial extension; (2) benign-appearing intracranial meningioma with extracranial metastasis; (3) originating from arachnoid cell rests of cranial nerve sheaths with extracranial growth; or (4) extracranial growth with no apparent connection with foramina or cranial nerve, presumably from embryonic rest cells.

According to the Hoye classification our case is an extracranial tumor that fits into either category 3 or 4. It is possible that the tumor may have arisen from arachnoid cell rests from the spinal nerve sheath, as the tumor was seen to insinuate into several intravertebral foramina, which would be category 3. It could also be category 4, as the tumor was shown not to be occluding the foramina.

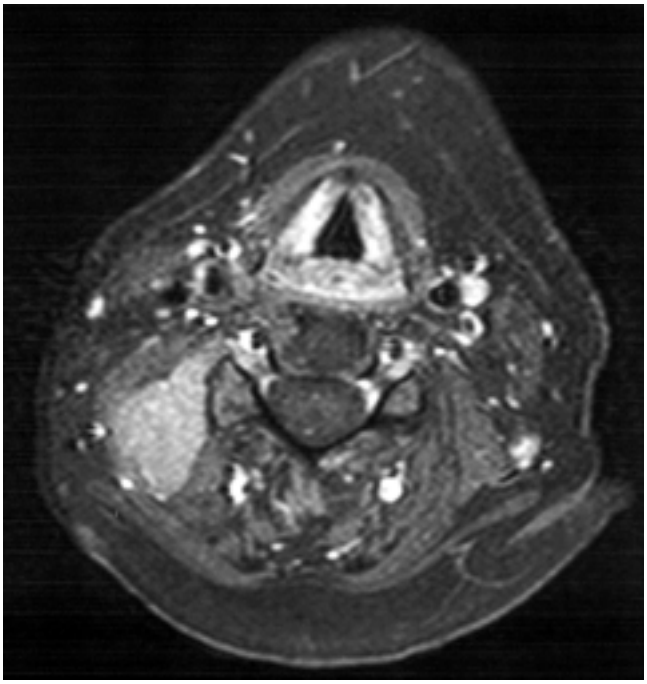

Figure 3. T1 axial post-gadolinium magnetic resonance image shows a mass that appears to be insinuating into several intravertebral foramina, but not occluding them.

Our patient presented with a rare case of neck swelling in the presence of another pathology in the cervical spine, and due to the gross neck fixation and curvature, the lump was masked for some time. The initial reaction by the patient and her clinician was that the lump was due to her AS. It is important therefore that the neck is examined carefully. Additional investigations including histopathology and imaging are needed to make an accurate diagnosis to exclude other causes of neck swelling.

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