СЛУЧАЙ ИЗ ПРАКТИКИ

EXTRA-AXIAL MENINGIOMA OF THE SKULL PRESENTING AS A SCALP MASS – REPORT OF A CASE AND REVIEW OF LITERATURE

S. Butt, P. O'Donnell, A. Saifuddin, I. Mahmood, A. Flanagan*

The Royal National Orthopaedic Hospital. The Departments of Radiology and *Pathology. London, UK

wellings in the scalp region are not uncommon. The usual differential diagnoses to be considered would include, a benign cyst (dermoid/ epidermoid or a trichilemmal cyst), an inflammatory lump, a haemangioma, osteoma, eosinophilic granuloma or a skull metastatic deposit. A meningioma is an unusual cause of this presentation. We report one such case and present a review of the relevant literature.

Keywords: swellings, scalp, meningioma, benign cyst, differential diagnoses.

ВНЕМОЗГОВАЯ МЕНИНГИОМА, ПРЕДСТАВЛЕННАЯ ОПУХОЛЬЮ МЯГКИХ ТКАНЕЙ СВОДА ЧЕРЕПА – КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ И ОБЗОР ЛИТЕРАТУРЫ

С. Батт, П. О'Доннелл, А.Саифуддин, И. Махмуд, А. Фланаган*

Королевский национальный госпиталь ортопедии. Кафедра лучевой диагностики и * патологии. Лондон, Великобритания

тек мягких тканей свода черепа встречается нечасто. Дифференциальный диагноз чаще всего включает в себя доброкачественные кисты (дермо-ид/эпидермоид или атерома), воспалительный процесс, гемангиома, остеома, эозинофильная гранулема или метастатическое поражение черепа. Довольно редко в таких случаях встречается менингиома. Нами представлен клинический случай и обзор литературы по этой теме.

Ключевые слова: отёк мягких тканей, свод черепа, менингиома, доброкачественные кисты, дифференциальный диагноз.

wellings in the scalp region are not uncommon. The usual differential diagnoses to be considered would include, a benign cyst (dermoid/ epidermoid or a trichilemmal cyst), an inflammatory lump, a haemangioma, osteoma, eosinophilic granuloma or a skull metastatic deposit. A meningioma is an unusual cause of this presentation. We report one such case and present a review of the relevant literature.

Case History.

A 70 year old man presented to a plastic surgeon for a Basal cell carcinoma behind his right ear. On examination, the doctor noticed an irregular swelling near the vertex of his skull. The swelling was firm in consistency and measured 3 cm by 2 cm in size. It was not fixed to the skin and was non-tender. On further questioning, the patient reported that this swelling had been present for about six years and was not growing in size. Radiographs of the area, (Fig. 1), showed the swelling to be causing erosion of the outer table of the skull and were of soft tissue density.

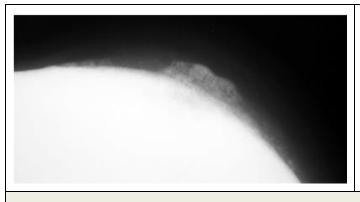
A CT of the area (Fig. 2) showed that the mass was deep to the scalp and was indeed causing erosion of the outer skull table.

No calcification or ossification was seen in the lesion. It was not shown to have an intracranial component or connection. The lesion showed significant enhancement after the administration of contrast medium.

A provisional diagnosis of a metastatic deposit was made and a needle biopsy was arranged. The biopsy showed the lesion to have spindle shaped cells of low to intermediate grade (Fig. 3).

After review of the slides by a panel of histopathologists specializing in neural diseases, the diagnosis of a meningioma without atypical features was made. The patient was a poor risk for general anaesthesia because of his cardiac status. Because of the benign nature of the mass, it was agreed to keep the lesion under observation and the patient was advised to have a follow-up MRI in three months time.

Discussion.



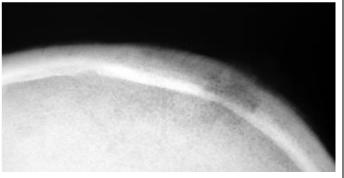


Fig.1. Plain radiographs of the skull showing erosion of the outer skull table with a soft tissue mass.

Scalp masses are seen quite commonly in patients of all age groups. In most cases the lump represents a benign condition and is incidentally noted. In children the common aetiologies for a scalp lump are dermoid cyst, epidermoid cyst, hemangiomas, hematomas and abscesses [1]. In the adult group trichilemmal cyst, inflammatory lesions, hemangiomas and lipomas are the usual causes. However, in a lump which is painful and/or is rapidly increasing in size, the possibility of a metastatic deposit should be considered. This is particularly true in a patient who has a known primary elsewhere.

In a major review of 388 scalp lumps, Carson et al [2] found 30 malignant lesions. 16 of these were metastatic deposits from a known primary in lungs, breast, kidney or thyroid. The rest were leukemia (three), lymphoma (four), melanoma (three), adenocarcinoma with unknown primary (two) and sarcomas (two). Bardales and Stanley [3] in their review of scalp lumps noted 9 lesions which were metastatic. They also showed five patients with metastasis who did not have a known primary elsewhere. Only two patients had a benign lump in their cohort of 16 patients who had been referred for a biopsy of a scalp mass.

Meningioma is a common tumour of the brain and spinal cord. In various studies they



Fig.2. CT of the skull, axial view, bone window shows the mass lesion causing skull vault erosion.

have been shown to represent 13-18% of all the primary intracranial tumors and up to 12% of all the primary intraspinal tumours [4,5]. The presentation of meningioma as a scalp lump is however uncommon.

These benign tumours arise from the arachnoid cells. Ectopic meningiomas are thought to arise from ectopic arachnoid cell rests or from pluripotential mesenchymal cells [6]. The ectopic arachnoid cell rests are formed because of delayed closure of the neural tube, with consequent herniation of the meninges or from premature closure of the neural tube with meningeal tissue being pinched off into the skin [7]. Arachnoid cells have been shown to be present in the diploic space of the skull. This explains the presence of ectopic meningiomas in the scalp, in the skull vault (the intra-osseous meningioma, where they are more commonly present along the sutures) and along the cranial nerves [8,9,10]. Intracranial meningiomas can extend outside the skull and malignant lesions can metastasize extra cranially [11,12]. Formation of ectopic meningiomas after trauma, which results in meningeal herniation outside the skull, has also been reported [7,13].

Pendergrass and Hope [14] described a meningioma in the frontal region which was causing erosion of the outer skull table without any intracranial connection. This was the first such case described in the radiology literature. Since then many reports of similar lesions have been published in the neurology and neurosurgery literature [28-31], but there have been very few reports in radiology journals. However, there have been reports of intradiploic/intraosseous meningiomas [8,16,17,18,19].

Unlike meningiomas related to the central neuraxis, ectopic meningiomas are rare. According to Farr et al [20] out of 405 cases of meningiomas only 3 were confirmed as primary extra cranial and extra spinal meningiomas present in the soft tissues. Wicker [11] and Waga [27] found the incidence of these ectopic meningiomas at 0.9% after reviewing 1768 and 226 cases respectively. Am-

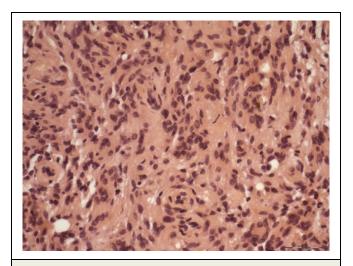


Fig.3. A transmitted light microscopic image of an haematoxylin and eosin-stained section showing a cellular lesion in which the cells have poorly defined borders and form the characteristic whorls around vessels or stromal elements. The finely distributed chromatin within the nuclei, with inconspicuous nucleoli, is characteristic of meningiomas.

mirati et al [21] found 4 intra diploic meningiomas in a series of 373 intra cranial lesions.

Apart from the scalp, meningiomas have been reported in the skin [7,23], orbit, paranasal sinuses, neck, nasal cavity and in the parotid glands [7,11,15,20].

Although commoner in adults, ectopic meningiomas occur at all ages and affect females more commonly than males [7,11,15,20].

Depending on their location, the symptoms range from proptosis, cranial nerve palsies, nasal blockage, or scalp/skin swelling.

It is important to be certain that the apparent scalp or sub-cutaneous tumour does not have an intra cranial component. Even when the lesion is extra cranial, there may be a rudimentary track, connecting the lump to the dura and if not careful, intervention can lead to meningitis or oth-

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er intracranial complication [7]. Moreover, an intracranial meningioma may have an extra cranial component. Farr et al [20] studied 405 cases of meningiomas and 71 of these tumors (20%) had extra cranial extensions. Fifteen of these patients had an ill-conceived extra cranial surgical intervention due to mis-diagnosis. In a review of the literature by Geoffray et al [15], out of 39 meningiomas of the naso-oral cavity and para nasal sinuses, 15 had documented intra cranial extension. In 84 extra cranial meningiomas 14 were present in the scalp with erosion of the outer skull table. None of these patients had any intra cranial extension.

Intra-diploic meningiomas can have osteolytic, mixed lytic and sclerotic or more rarely completely osteolytic appearance on plain x-rays [9]. Osteoblastic response is said to be secondary to the infiltration of meningeal cells in the bone resulting in osteoblastic activity (19). Sclerosis can affect the diploic space or one or both of the skull tables [8]. In most cases there is inward bulging of the inner skull table. Lesions on the inner or outer aspects of skull can cause erosion of the adjacent skull table. Calcification is better seen on CT [23,24,26]. MR shows a low signal mass on T1W, which is high signal on T2W, and after gadolinium administration, the intraosseous lesion enhances in a spotty manner and the extra cranial component enhances uniformly [23,24,26].

The diagnostic work-up of a scalp lump should include a CT which helps to show the skull erosion and the nature of the soft tissue. MR shows any intracranial extension. Biopsy is needed to establish a tissue diagnosis and is most easily done under ultrasound guidance.

In conclusion an ectopic meningioma should be considered in the differential diagnosis of a scalp lump. It is important to investigate for an intracranial extension of an apparent extra cranial mass lesion to avoid a potentially serious complication.

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RUSSIAN ELECTRONIC JOURNAL OF RADIOLOGY

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