International Journal of Gerontology 9 (2015) 183–186

Contents lists available at ScienceDirect

International Journal of Gerontology

journal homepage: www.ijge-online.com

Case Report

Malignant Primary Extradural Meningioma in Elderly Patients: Report of Two Cases and Literature Review☆

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ARTICLE INFO

Article history:
Received 2 December 2013
Received in revised form
24 January 2014
Accepted 25 June 2014
Available online 3 September 2015

Keywords:
ectopic meningioma,
primary extradural meningioma,
primary intraosseous meningioma

SUMMARY

Primary extradural meningioma, also known as ectopic meningioma, is uncommon and represents ~2% of all meningiomas. Malignant primary extradural meningioma is even more rare. The rarity of cases means the condition is not readily suspected or diagnosed. In this report, we present one case of primary extradural meningioma in elderly patients who presented with a scalp nodule and another highly suspected case. Both patients underwent surgical resection of the tumors. Histological study and immunostaining confirmed the diagnosis. We also conducted a literature review regarding similar cases. Surgical resection with a wide margin remains the first choice of treatment.

1. Introduction

Meningioma constitutes 14.3–19% of all primary intracranial neoplasms[1]. However, primary extradural meningioma (PEM), also known as ectopic meningioma, is rarely reported in fewer than 2% of the meningiomas[2–4]. Extradural meningiomas have histological features similar to meningioma of the central nervous system and are generally benign. It made malignant ectopic meningioma even rarer. Here we report two cases of ectopic meningioma with World Health Organization (WHO) Grade III and review some of the literature.

2. Case reports

2.1. Case 1

An 80-year-old woman presented with a 2-year history of a lump overlying the right parietal region of the scalp. The mass lesion was noted after head trauma 2 years earlier, and local treatment with aspiration of bloody fluid was performed at that time. She visited our clinic 1 year ago, and a skull radiograph (Figure 1) showed a soft-tissue density ~2 cm in height. The patient returned to our clinic because of rapid growth of the scalp mass, which became 5 cm thick and 6 cm in diameter (Figure 2) without any changes in the overlying skin. The lump was firm, smooth, and not tender. Neurological examination revealed no focal neurological deficit and the patient also denied headache, dizziness, or nausea/vomiting. A computed tomography (CT) scan (Figure 3) demonstrated a right parietal subcutaneous mass lesion ~6 cm in diameter with bony destruction and extension to the superior sagittal sinus. The first impression was skin malignancy or metastatic carcinoma, judging by the aggressive behavior of the tumor, and wide surgical resection was planned. During surgery, the tumor appeared soft and reddish with hypervascularity and was located just beneath the skin with dura invasion. Near-total resection of the tumor was achieved. We left some residual tumor close to the superior sagittal sinus. Immunohistological examination revealed a hypercellular tumor composed of pleomorphic epithelioid or short-spindle cells with brisk mitosis and focal necrosis. Whorl formation was also noted. The tumor cells showed positivity for vimentin (diffuse +), epithelial membrane antigen (EMA; focal +), cytokeratin (CK; AE1/AE3; focal +), and P63(focal +). The staining results of S-100 protein, CK5/6, CK7, and CK20 were all negative. The findings were compatible with anaplastic (malignant) meningioma, WHO Grade III (Figure 4). Although preoperative image was limited, on review of the skull film and clinical progression, the tumor was highly suspected to be initially located subcutaneously and invaded

☆ Conflicts of interest: All contributing authors declare that they have no conflicts of interest.

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http://dx.doi.org/10.1016/j.ijge.2014.06.006

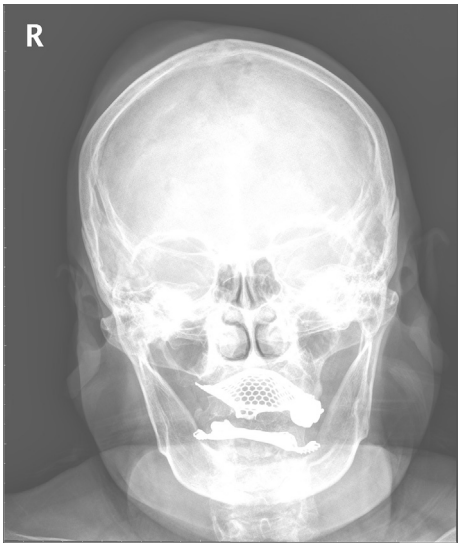

Figure 1. Skull radiograph showing a soft-tissue density at the right parietal region.

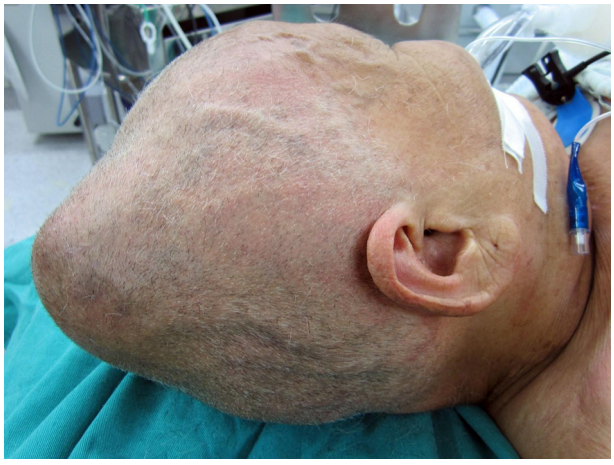

Figure 2. Gross appearance of the lump at the right parietal area after preoperative shaving.

intracranially. The final diagnosis was cutaneous meningioma with intracranial extension. The postoperative course was uneventful and radiotherapy was indicated because of the diagnosis of malignant meningioma. However, the patient's family refused radiotherapy because of the risks of complication.

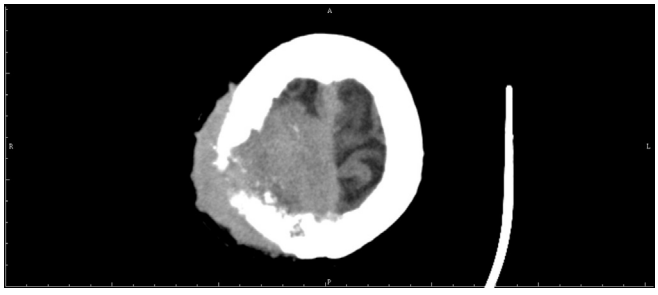

Figure 3. Brain CT scan showing tumor caused by bony erosion.

2.2. Case 2

An 81-year-old woman visited our clinic because of the presence of a lump on the right frontal region of the skull for several months. The mass was firm and fixed with a diameter of ~5 cm. Skull radiograph and brain CT scan revealed an osteolytic lesion of the right frontal bone. Brain magnetic resonance imaging (MRI) showed a mass lesion at the right frontal skull with bony destruction and intracranial invasion with attachment only to the dura mater (Figure 5). En bloc resection of the tumor with a 0.5-cm margin was performed through a right frontal craniectomy. The tumor was only attached to the dura and there was dural infiltration. Immunohistological examination showed that the tumor was composed of epithelioid and spindle cells, and foci of necrosis and hemorrhage were easily found. The tumor showed immunoreactivity for vimentin and CD31 and indicated occasional positive staining for Cytokeratin but not for CD34, friend leukemia integration-1, factor VIII, EMA, or glial fibrillary acidic protein. The findings were compatible with anaplastic (malignant) meningioma, WHO Grade III. The final diagnosis was primary intraosseous meningioma with intracranial extension.

3. Discussion

PEM refers to any meningioma arising outside the intracranial compartment. A variety of locations were reported, including the calvarium[5], orbit, middle ear, paranasal sinuses, neck, skin, lung, and mediastinum[6], etc. A review of 162 reported cases by Mattox et al[7] demonstrated that 85.4% of PEMs are found in the head and neck region. Here we reported two cases with different types of PEMs and both presented with a scalp lump. The first was a case of cutaneous meningioma, in which tumor is derived from ectopic meningoepithelial cells in the dermis or subcutaneous tissue[8]. Most are located within the scalp[9] and are also known as scalp meningiomas. The other case is primary intraosseous meningioma (PIM). Although Hoye et al[10] defined PIMs as having no dura involvement[9], Oka et al[11] reported that some calvarial meningiomas infiltrated dura but not primary intradural origin. Lang et al[2] also agreed that dural invasion is not an exclusion criterion for PIM, but location and growth pattern are more important. In our first case, initial imaging studies showed intact bony structure with a subcutaneous soft tissue lesion; however, with disease progression, the tumor invaded the intracranial compartment. Final pathological diagnosis in both cases was anaplastic meningioma; this may explain why both cases showed rapid growth and aggressive behavior with intracranial invasion.

Patients with skull or scalp PEM mostly present with a painless mass with slowing growth for months to years[7]. Moreover, according to their size, location, and relation to the cranial nerves, symptoms including focal neurological deficits, seizure, tinnitus, headache, or dizziness may also occur. In our report, both cases presented with a painless mass but the lesions showed relatively rapid growth and aggressive behavior.

PEMs occur mostly in older adults with a median age at diagnosis in the fifth decade[7]. In our report, both patients were both older 80 years old. They searched for medical help only when the PEMs were large enough to cause symptoms.

Radiological findings are variable. Most PEMs are osteoblastic lesions, but osteolytic type may be found in <20% of cases[5,12,13]. Noncontrast CT scans in osteolytic lesions revealed hypodense lesions causing thinning, expansion, and even destruction of inner or outer cortical layers of the bone. MRI showed hypointense lesions in T1-weighted images and hyperintense in T2-weighted MRI with strong gadolinium enhancement. MRI images also help evaluate the relationship with dura and any intracranial extension.