Intradiploic Meningioma Mimicking Calvarial Metastasis: Case Report

Kalvarial Metastazı Taklit Eden İntradiploik Meningioma: Olgu Sunumu

ABSTRACT

Meningiomas are the most common benign intracranial neoplasms. Nearly 20% of all primary intracranial tumors are meningiomas. Primary intraosseous meningiomas are a subtype of the meningiomas that represents the most uncommon manifestation of meningiomas. Although rare, these tumors can be found to occur in unexpected areas of the head and neck. The patient was a 78-year-old male who was operated two times for urinary bladder cancer. During his routine oncology follow-ups, the PET scan demonstrated a hyperactive area in the right parietal bone. Preoperative diagnosis was a metastasis, but histological examination revealed an osteolytic interosseous meningioma. The possibility of an intraosseous meningioma mimicking a metastatic tumor should be kept in mind.

KEY WORDS: Skull, İntradiploic, Meningioma, Metastasis

ÖZ

Meningiomlar en sık görülen iyi huylu intrakranial tümörlerdendir. Tüm primer intrakranial tümörlerin yaklaşık % 20' sini meningiomlar oluşturur. Primer intraosseöz meningiomlar ise meningiomların çok nadir görülen alt gruplarından biridir. Mesane tümörü nedeniyle daha önce 2 defa opere edilmiş olan 78 yaşındaki erkek hasta tarafımıza rutin malignite kontrollerinde yapılan PET taramasında parietal kemikte metastaz ile uyumlu hiperaktif alan saptanması sonucu başvurmuştur. Ameliyat öncesi tanı metastaz olmasına rağmen histopatolojik tanı operasyon sonrası meningiom olarak bildirildi. Intraosseöz meningiomların metastazları taklit edebilecekleri akıldan çıkartılmamalı ve ayırıcı tanıda akılda tutulmaları gerekir.

ANAHTAR SOZCÜKLER: Kafatası, Kemik içi, Meningioma, Metastaz

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INTRODUCTION

Although intraosseous meningiomas are very rare, they are very well described in the literature. Extradural meningiomas arise in locations other than the dura mater like neck, nasopharnyx or skin. Winkler, in 1904, first described a meningioma originating in an extradural location (24). These lesions can be confused with metastatic carcinomas and bone tumors. Here we report a case of intradiploic meningothelial meningioma that was active on the PET scan and mimicked metastasis.

CASE REPORT

A 78-year-old man was operated for urinary bladder cancer two times in 2004 and 2007. PET scan showed a hypermetabolic area in the right parietal bone (Figure 1) during follow-up for the malignancy. Clinical examination revealed a 2 x 2 x 1 cm domeshaped slight bony protuberance in the same area. The lesion was bone hard and the overlying skin demonstrated no notable alterations and was not attached to skin. The neurological examination showed no deficits. Plain skull radiographs revealed 2 x 1 cm low-density area in the right parietal bone (Figure 2). There was no history of any trauma. Hematological and biochemical investigation results were within the normal range. The lesion was further characterized by using pre- and post-contrast MR imaging. The lesion was hypointense on T1weighted and hyperintense on T2-weighted images. The underlying dura and brain were normal. The lesion showed homogenous enhancement following gadolinium injection (Figure 3). During surgery, the external tabula of the cranium was carefully drilled using a high-speed pneumatic drill. The lesion was well demarcated from the calvarium and placed in



Figure 2: Lateral skull X-ray film showing the osteolytic area in the right parietal bone.

an antrum between the two tabulae of the skull. Tumor was totally intradiploic in location. Total resection was performed. The inner table of the cranium was intact and was not opened. Histopathological examination of the specimen confirmed the presence of meningothelial meningioma (Figure 4). The postoperative period was uneventful.

DISCUSSION

Meningiomas occur with an estimated incidence of 2.1 per 100,000 cases. They typically appear in patients 20 to 60 years of age (peak incidence at 45



Figure 1: PET-CT scan demonstrating the hypermetabolic area in the right parietal bone.



Figure 3: (Left) Axial, (Middle) Coronal, (Right) Sagittal T1-weighted MR images after Gd contrast administration. The mass enhances homogenously in the right parietal bone.



Figure 4: Diffuse meningothelial proliferation without any evidence of atypia, necrosis, sheeting, hypercellularity, prominent nucleoli and small cells. Syncytial cells compatible with meningothelial meningioma are the most striking finding (H&E, original magnification x100).

years) with a female-to-male ratio of approximately 2:1 (6,19). Intraosseous meningiomas constitute only 2% of all meningiomas (21).

'Primary intraosseous meningioma' is a subset of extradural meningiomas that arise in bone (23). True primary intraosseous meningiomas do not involve the underlying dura. Extradural meningiomas that arise from the skull have been named as calvarial, intradiploic, and intraosseous (7). To avoid confusion, Lang et al. classified interosseous meningiomas as purely extracalvarial (type I), purely calvarial (type II), or calvarial with extracalvarial extension. The latter two are further divided as convexity (C) or skull base (B) forms (17). The real incidence of primary intraosseous meningioma is unknown but they have been largely reported in the literature. The average age was 50.5 with a slight female predominance 1.65:1 (18). Convexity and the skull base are the two major locations for intraosseous meningiomas (4,7,11,12,16).

The origin of these tumors is controversial. Some authors believe they originate from arachnoid cap cells in normal dura and in the arachnoid granulations. Others have mentioned that they can arise from ectopic meningocytes trapped in the cranial sutures during molding of the head at birth or following blunt trauma (3). A few recently reported cases had a history of trauma to the head and a subsequent fracture line near the region of the intraosseous meningioma (8). There were no suspected etiological factors in our patient.

Neurological signs and symptoms are usually absent in patients. The initial symptom is usually a painless expansive mass with normal neurological findings. The symptoms are dependent on tumor location, size, and involvement of the neighboring structures; presenting symptoms such as neurological deficit, dizziness, hearing loss, and seizures have also been reported in the literature.

Meningiomas presenting with scalp swelling and extracranial soft-tissue masses are more aggressive in nature than others (20). Osteolytic meningiomas associated with a soft-tissue component must be considered malignant until proven otherwise (22).

Radiographic findings of EM are limited and not pathognomonic because of the superimposed bony

structures (10). The tumors are typically either osteoblastic or osteolytic, although mixed versions have been reported. Plain X-ray skull radiographs can detect abnormalities, especially in the osteoblastic type. Hyperostosis, irregular foci of calcification, and atypical vascular marking can be seen. The osteolytic type can be distinguished by its hypodense appearance on plain radiographs as in our case. T1-weighted images display isodense or hypodense lesions compared with the brain while T2-weighted images are variable but usually hyperintense on magnetic resonance imaging. Prominent homogeneous enhancement after Gadolinium (Gd) administration is typical. Normally the lesions do not show a 'dural tail'. If the dural tail appearance is seen, it could be secondary to dural irritation or invasion by the tumor (2). In our case, the lesion was hypointense on T1-weighed images and enhanced homogenously after Gd enhancement without dural tail appearance. Angiography is useful when embolisation is required although it is reserved for intracranial meningiomas (4).

Histopathologically, these lesions may range from epithelial to mesenchymal in appearance like their intracranial counterparts (5, 10).psammoma Microscopically, bodies and eosinophilic tumor cells with distinct borders in clusters and whorls can be seen (7,13). The nuclei are typically oval and regular and nuclear pseudoinclusions may be observed. Although they are benign lesions, recent studies emphasize that intraosseous meningiomas have a higher incidence of malignant characteristics compared to intradural meningiomas (22,27). The typical findings were found to be compatible with a meningothelial meningioma in our case without any resemblance to a normal or giant arachoidal granulation. By definition, arachnoid granulations (AGs) or Pacchionian bodies consist of evaginations of the pia-arachnoid into the dural sinuses or lacunae laterales; cerebrospinal fluid (CSF) drains primarily through these structures, and as such, their malfunction may underlie some cases of communicating hydrocephalus (1). By and large these granulations are composed of numerous arachnoid villi; these villi have a thin outer limiting membrane (composed of arachnoid mesothelium and vascular endothelium, demonstrating partial fusion), and a central core composed of collagenous and elastic fibers intermingled with a tubular network (1,2,3). These tubules are in continuity with the subarachnoid space.

The differential diagnosis of the osteoblastic type must include osteoma, osteosarcoma, Paget's disease, meningioma en plaque and fibrous dysplasia (9,14). The intradiploic osteolytic subtype is an extremely rare tumor and only 16 cases have been reported in the literature so far (25). The osteolytic subtype must be differentiated from hemangioma, chondroma, chondrosarcoma, dermoid, epidermoid tumor, brown tumor, multiple myeloma, plasmacytoma, giant cell tumor, aneurysmal bone cyst, eosinophilic granuloma, or metastatic cancer (1,8,15,26).

The only curative treatment modality is complete resection. Cranial reconstruction must be done if the surgical resection is wide. When the tumor is resected subtotally because of the involvement of critical structures, the residual tumor should be followed up radiologically. Radiation therapy is an option for patients with progressive residual tumor (8). Other adjuvant treatment modalities may be chemotherapy and bisphosphonate therapy.

In conclusion, although rare, osteolytic primary intraosseous meningioma should be considered in differential diagnosis of calvarial lesions even if the lesion seems hypermetabolic on PET scan and is diagnosed as metastasis. We recommend removing the single skull mass in patients with known cancer.

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