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Intraosseous Meningioma Mimicking Osteosarcoma in an Adolescent: A Case Report

Bir Adolesanda Osterosarkomu Taklit Eden İntraosseoz Menenjiom: Olgu Sunumu

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ABSTRACT

Intaosseous meningiomas (IM) are the one of the less frequent benign tumors of the skull. The etiology of IM has not been cleared yet. The frontoparietal and orbital regions are the most common locations for IM. The average age for IM diagnosis is 50.5. A 16-year-old girl with a right frontoparietal mass was referred to our outpatient clinic. Cranial CT revealed a mass lesion which resulted in expansion in the right parietal and posterior frontal bone, having lytic and sclerotic regions inside with accompanying irregular cortex in inner and outer tables of the calvarium. Prediagnosis was osteosarcoma according to the imaging studies and after the performed biopsy and consecutive surgery, the lesion was diagnosed as IM. Though CT with bone windows is often useful, it is not always diagnostic. Biopsy plays an important role in calvarial vault lesions for planning the treatment. In our case, malignant criteria in radiology did not match the benign histology revealed. Radiological preoperative misdiagnosis of meningioma is possible.

KEYWORDS: Intraosseous meningioma, Osteosarcoma, Adolescent

ÖZ

Intraosseöz menenjiomlar(IM) kafatasının en az görülen tümörlerindendir. Etiyolojisi hala aydınlatılamamıştır. Intraosseöz menenjiomlar için frontoparyetal ve orbital bölgeler en sık lokalizasyonlardır. Başında yaklaşık 2 aydır varolan ağrısız ve sert şişlik şikayeti ile tarafımıza başvuran 16 yaşındaki kız hastanın istenen kraniyal bilgisayarlı tomografisinde sağ paryetal ve posterior frontal kemikte içinde litik ve sklerotik alanlar içeren, ekspansiyona yol açmış, iç ve dış tabulada irregüler kortikal yapıyla seyreden kitlesel lezyon saptandı. Lezyonun ön tanısı radyolojik olarak osteosarkoma idi ve biyopsi sonrası intraosseöz menenjiom lehine sonuç gelmesi üzerine total rezeksiyon yapıldı. Kemik pencereli bilgisayarlı tomografi görüntüleri tanı için genellikle faydalı olmalarına rağmen, her zaman tanısal değildir. Biyopsinin kalvaryal lezyonların tedavisini planlamakta önemli bir rolü vardır. Bu olguda radyolojideki malign kriterler benign histolojiyle uyuşmamıştır. Menenjiomalar, preoperatif yanlış tanı alabilen tümörlerdir.

ANAHTAR SÖZCÜKLER: İntraosseoz menenjiom, Osteosarkom, Adolesans

INTRODUCTION

Meningiomas are the most common benign intracranial neoplasms. Primary intraosseous meningioma (IM) is a subtype of the extracranial form that likely represents the rarest manifestation of meningiomas. They are mostly seen in adults. McGuire et al, in their analysis of the literature, have stated that the average reported age presenting with extracranial IMs was 50.5 (ranging from 10 to 80 years) (9). According to the cases included in this review, there had been only four patients reported younger than 18 years old and their tumor locations were frontoparietal, frontal, frontoorbitonasal and temporal bone.

We report a 16-year-old girl who had a right frontoparietal mass and underwent surgery, and was diagnosed as IM.

CASE REPORT

A 16-year-old girl with a right frontoparietal mass was referred to our outpatient clinic. Her chief complaint was the mass localized on right side of her head. She had noticed the hard, painless mass for about two months and had no other complaints other than esthetic consideration. She had no trauma history or a past medical history. On physical examination a 4x5 cm bony protuberance which was hard, non-tender and painless in origin, was observed on right frontoparietal region. It was not attached to the skin, and there were no skin changes on examination. Her remaining physical examination was normal, and no neurological deficits were found on admission. Blood tests, including full blood count, electrolytes, biochemical parameters, erythrocytes sedimentation rate were all in normal limits.

Cranial CT revealed a mass lesion which resulted in expansion in the right parietal and posterior frontal bone, and included lytic and sclerotic regions inside with irregular cortex in inner and outer tables of the calvarium. The medial part of the mass had a relation with adjacent soft tissue with mild compression of the frontal lobe and obliteration in the sulcus.

The right lateral ventricle had a partial obliteration and a 4 mm right to left shift was noted (Figure 1A,B). Post-contrast cranial MRI showed heterogeneous contrast enhancement and noticed thickness and contrast enhancement in adjacent dura underlying the right frontoparietal region (Figure 1C). The radiological preoperative diagnosis was osteosarcoma as the mass lesion with expansion in right frontoparietal region had malign radiological criteria.

A bone biopsy was performed and pathology revealed IM of the skull. After the biopsy, we planned a total resection. We performed a frontoparietal craniectomy under general anesthesia. After reflecting the scalp, haemorrhagic tumor was noticed in the outer tables of the right parietal bone. A right frontoparietal craniectomy was performed (Figure 2B). The tumor seemed to originate from the diploic space and had expanded mainly inside but also outside. Underlying dura was infiltrated. After the invaded dura was removed, the dural defect was patched with an otologous fascia lata graft. Cranioplasty with methyl methacrylate was performed. The postoperative course was uneventful. Histological examination showed meningiamotous infiltration between bone trabeculae and accompanying active chronic nonspecific inflammation. Meningioma was reported to be the transitional type (WHO Grade 1). The tumor showed medium hypercellularity and pleomorphism (Figure 2A). Mitotic index of Ki 67 was 3-4 % and 5-10 % of the cells had progesterone receptor positivity.

DISCUSSION

The etiology of IM has not been cleared yet. However, proposed theories include some presumed ectopic arachnoid cap cells in an extradural location, entrapment/detachment of displaced pacchionian bodies during embryonic development, displacement of arachnoid islets by a traumatic event or cerebral hypertension or a separate origin from

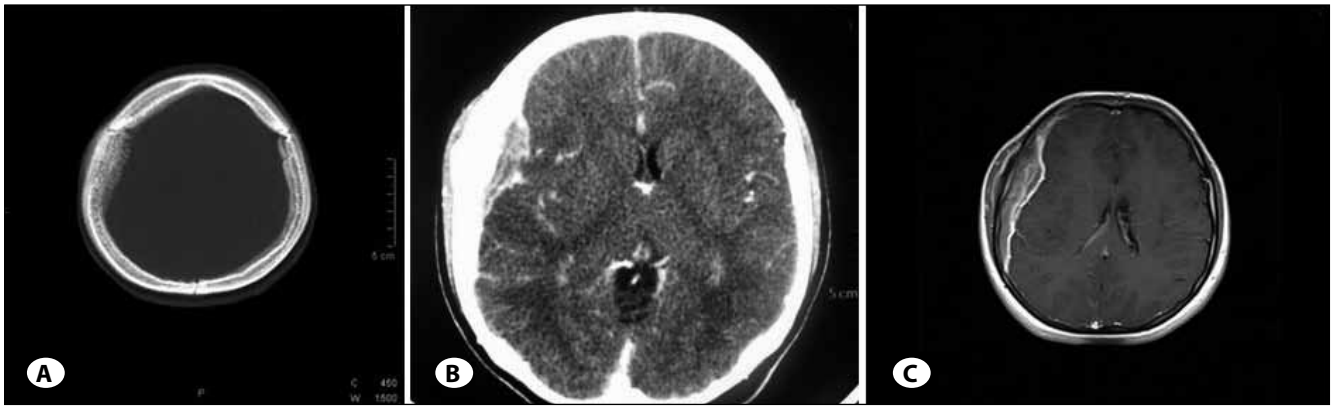

Figure 1: A) Axial-bone window CCT imaging showing right frontoparietal mass with lytic and sclerotic region inside with irregular cortex in inner and outer tables of the calvarium. **B)** Axial contrast-enhanced CCT image demonstrates the contrast enhancement in the underlying dura and the medial part of the mass had a relation with adjacent soft tissue with mild compression of the frontal lobe and obliteration in the sulcus. **C)** Post contrast axial T-1 weighted cranial MRI shows heterogeneous contrast enhancement and noticed thickness and contrast enhancement in adjacent dura underlying the right frontoparietal region.

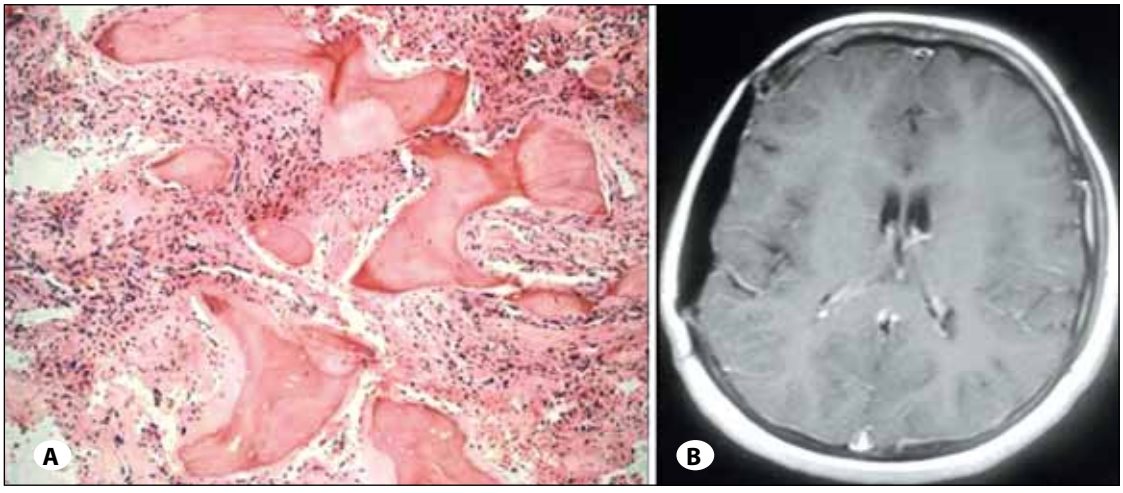

Figure 2: A) Left Photomicrograph of the tumor showing meningioma infiltration between bone trabecules and accompanying active chronic nonspecific inflammation, continuing inside the adjacent striped muscle. HE stain x 100. **B)** Post-operative post contrast axial T-1 weighted MRI image showing right frontoparietal craniectomy and postoperative changes.

a differentiated or multipotential mesenchymal cell (11). Wrinkler, in 1904, first described a meningioma originating in an extradural location (4). The true incidence of primary meningiomas is unknown as they have largely been reported in the literature as case reports (8). True primary IM is defined as a lesion that does not involve the underlying dura (3, 12). However, IM scan be classified as primary and secondary, and the dura may become involved later in the course of the tumor growth even in primary IM (1). Secondary IM is due to the extension of an intracranial meningioma into the calvarium (1). However, the underlying dura may be affected by primary or secondary IMs (6).

Lang and colleagues have suggested a classification for primary extradural meningiomas. Tumors that are purely extracalvarial are Type I, purely calvarial tumors are Type II, and calvarial tumors with extracalvarial extension are Type III. Each category is further divided into convexity (C) or skull base (B) subtypes based on their anatomical location. Thus, IMs could be considered Type II or III primary extradural meningiomas based on whether extracalvarial extension is observed (7). Bassiouni et al have modified this classification system, particularly considering involvement of the dura mater by the tumor process and suggested as location between outer and inner calvarial table-epidural as Type 1, location between outer and inner calvarial table-diploic as Type 2, extracalvarial location outside outer calvarial table as Type 3, and extension from dura to extracalvarial space as Type 4 (2).

According to the literature, 68% of the primary extradural meningiomas involved the calvaria. Frontoparietal and orbital regions are the most common locations for IMs. Other sites reported in the literature are subcutaneous tissue of the skin, paranasal sinuses, nasal cavities, oral cavities, parapharyngeal space, neck, salivary glands, and along the perineural sheath of the cranial nerves (13).

The radiographic differential diagnosis is case dependent and often vague, as these tumors can often be mistaken for a number of other osseous lesions, including osteosarcoma, bone metastases, fibrous dysplasia, Paget's disease, ossifying fibroma, and osteoma (9). The differential diagnosis for a primary lytic calvarial lesion includes lytic metastasis, epidermoid tumor, multiple myeloma, eosinophilic granuloma, fibrous dysplasia, Brown tumor, skull dermoid, lytic intraosseous meningioma, giant cell tumor, hemangiopericytoma, and hemangioma. Though CT with bone windows is often useful, it is not always diagnostic. Radiological preoperative misdiagnosis of meningioma is possible (8, 10). The characteristic MR imaging appearance of the intraosseous meningioma, demonstrating homogeneously dense Gd enhancement of the tumor within the skull, may help distinguish this lesion from meningioma en plaque, as well as from other osteoblastic skull lesions such as osteoma (nonenhancing), osteosarcoma (irregular contours, heterogeneous signal, and enhancement) and Paget disease (heterogeneous signal, nonenhancing) (5). Likewise, the histological differential diagnosis may also be vast and can include osteosarcoma, hemangiopericytoma, and soft tissue spindle cell tumors—especially those of peripheral nerve origin (2). Biopsy plays an important role in calvarial vault lesions for planning the treatment. In our case, malignant criteria in radiology did not match the benign histology revealed.

In the review of McGuire et al, there has been limited number of case studies reporting patients under 20 year old having IM (9) and the localizations were frontal bone, frontoorbitonasal, frontoparietal bone, and temporal bone. Our patient was one of the rare young patients diagnosed to have this rare tumor.

Maximal tumor resection and cranial remodeling are important aspects of treatment for intraosseous meningiomas. Adjuvant therapy may be considered in cases in which patients have unresectable tumors causing neurological deficit or demonstrating malignant or atypical features histologically (5).

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