



Mount Meningioma with Tumor Cap

Tümör Kapaklı Tepecik Menenjiyomu

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ABSTRACT

The association between meningioma and hyperostosis of adjacent calvarium is well established. However, the cause of hyperostosis is still not certain. Various authors in the past have tried to explain this phenomenon by preceding trauma or by bone irritation by the tumor without bone invasion, while some believe it to be because of stimulation of osteoblasts in normal bone by factors secreted by tumor cells. There are still others who believe it to be secondary to production of bone by the tumor itself, while some have hypothesised that vascular disturbances secondary to the tumor result in hyperostosis. The authors wish to report a case of parasagittal meningioma in which the bone overlying the meningioma was so hyperostotic that it appeared to look like a 'mountain'. In addition, the tumor appeared to invade through the bone to form a 'tumor cap' over the hyperostotic bone.

KEYWORDS: Meningioma, Bone changes, Hyperostosis, Mount meningioma, Bone invasion, Tumor cap

ÖZ

Menenjiyom ve komşu kalvarium hiperostozu arasındaki ilişki iyi belirlenmiştir. Ancak hiperostozun nedeni kesin değildir. Geçmişte çeşitli yazarlar bu olayı kemik invazyonu olmadan tümör nedeniyle kemik tahrişi veya önceki travmayla açıklamaya çalışmışken diğerleri ise tümör hücrelerinin salgıladığı faktörlerle normal kemiklerde osteoblast stimülasyonu nedeniyle oluştuğuna inanmaktadır. Ayrıca başka uzmanlar tümörün kendisinin kemik üretimine sekonder olduğunu ve yine başkaları tümöre sekonder vasküler bozuklukların hiperostozla sonuçlandığı kanısındadır. Yazarlar menenjiyom üzerindeki kemikteki hiperostoz derecesinin bir 'dağ' gibi gözükmesine yol açtığı bir parasagittal menenjiyom olgusu bildirmektedir. Ayrıca tümör, hiperostotik kemik üzerinde bir 'tümör kapağı' oluşturacak şekilde invazyon yapmış görünümdeydi.

ANAHTAR SÖZCÜKLER: Menenjiyom, Kemik değişiklikleri, Hiperostoz, Tepecik menenjiyomu, Kemik invazyonu, Tümör kapağı

INTRODUCTION

Meningioma is known to cause various bone changes of the overlying calvarium in the form of erosion, hyperostosis or blistering, enlargement of vascular grooves (especially middle meningeal artery) or sinus pneumatosis dilatans. The association between meningioma and hyperostosis is well established (1, 11). It is more frequently seen in the meningiomas involving the convexities and sphenoid wing (11).

The authors report an interesting case of parasagittal meningioma in which the bone overlying the meningioma was so hyperostotic that it appeared to look like a 'mountain'. Furthermore, the tumor appeared to invade through the bone to form a 'tumor cap' over the hyperostotic bone. To the best of our knowledge, such severity of hyperostosis along with tumor cap is extremely rare.

CASE REPORT

A 17-year-old girl presented to us with a bony hard, painless, progressive swelling at the top of her head for five years. The patient was a known case of neurofibromatosis type 2. Examination revealed a 7 cm x 5 cm x 4 cm sized, bony hard,

nontender swelling at the vertex, more towards the right side. The swelling had smooth margins and healthy overlying skin.

CT scan of the head revealed three mass lesions, one each in the bilateral CPA cisterns and one in the parasagittal region. The bone overlying the parasagittal region mass lesion revealed gross hyperostosis (Figure 1A-D). The MRI scan of the patient revealed a 6.6 x 3.8 x 6.9 cm sized heterogeneously enhancing mass lesion along the superior calvarium in the middle third parasagittal region with involvement of the superior sagittal sinus. In addition, the tumor appeared to invade through the bone to form a 'tumor cap' over the hyperostotic bone (Figure 1A-D). A diagnosis of NF2 with bilateral vestibular schwannomas and parasagittal meningioma was made. The patient was planned for right frontoparietal craniectomy and gross total excision of the right parasagittal meningioma. In view of the expected hypervascularity of the skin and bone, deep suture bites were taken all around the planned incision to reduce the blood supply to the scalp overlying the tumor. Intraoperatively, the bone was found to be severely hyperostotic and the meningioma mass appeared to have invaded through the bone and was present beneath the scalp above the hyperostotic bone, forming a 'tumor cap'. The

extracranial mass was decompressed using loop cautery. A bone flap was raised using Midas Rex B1 drill after making burr holes. The dura was badly adherent to the bone and appeared infiltrated by the tumor. The tumor was grayish white, soft, highly vascular, CUSA suckable, right parasagittal in location and involved the sino-dural angle. Simpson excision grade III was done in order to avoid injury to the superior sagittal sinus. The hyperostotic bone around the craniotomy was drilled with Midas Rex M8 drill bit after lax duraplasty. The post-operative CT scan revealed hydrocephalus and accordingly left-sided low-pressure ventriculo-peritoneal shunt procedure was

performed. She made good recovery in the post-operative period.

The histopathological examination revealed a transitional meningioma (WHO grade I) with a MIB labeling index of approximately 6%. The biopsy taken from the hyperostotic bone revealed tumor invasion into the bony trabeculae (Figure 2A-D).

The patient was ambulatory and doing well at the last follow up 3 weeks after discharge and is planned for management of bilateral vestibular schwannomas followed by a period of 3 months.

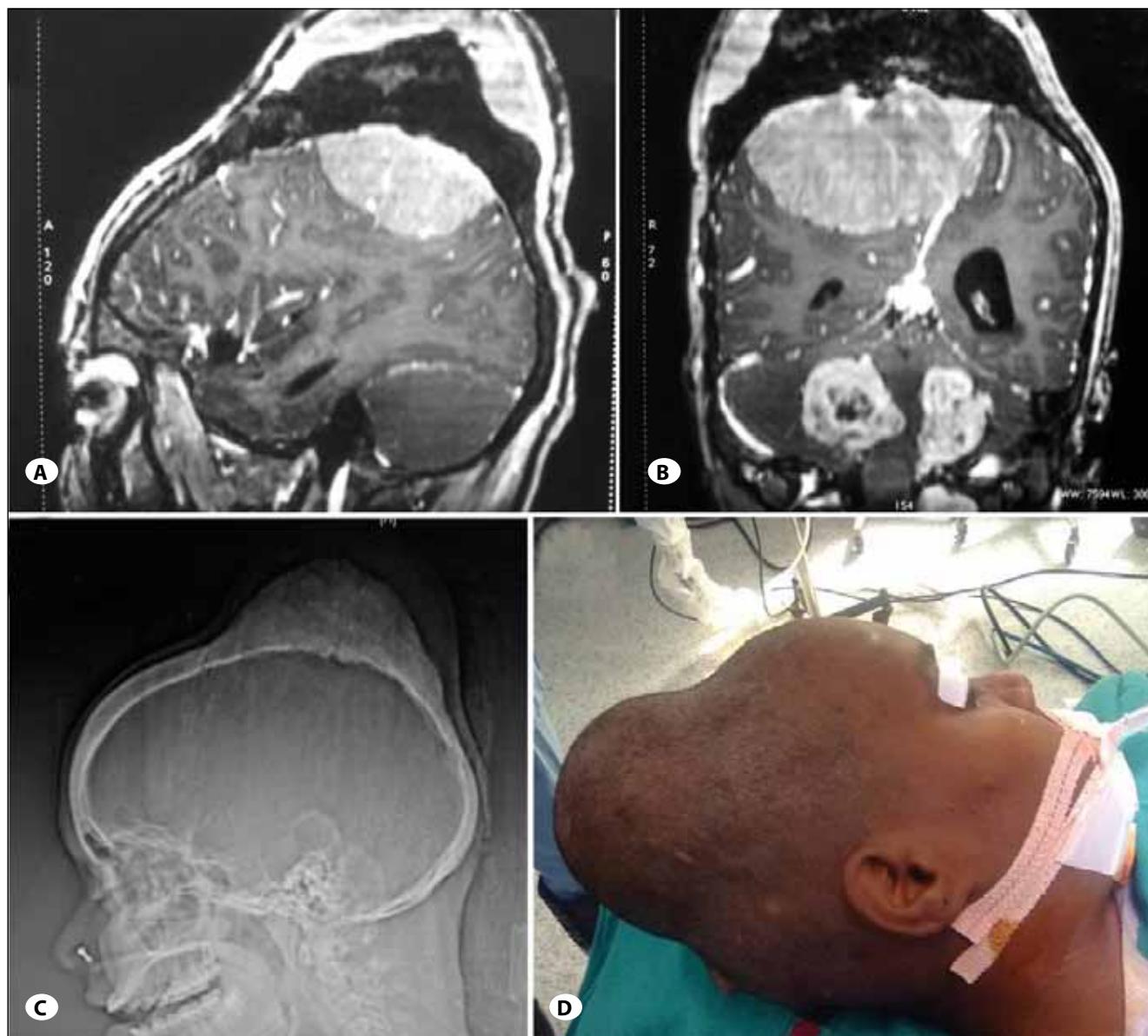


Figure 1: A) Sagittal and B) coronal sections of preoperative contrast enhanced Magnetic resonance imaging of the patient revealing a middle third parasagittal meningioma with gross hyperostosis and tumor cap overlying the hyperostotic bone. The main tumor mass and the tumor cap over the hyperostotic bone is brilliantly enhancing on contrast administration. Coronal image (b) also shows bilateral vestibular schwannomas; C) CT scannogram of the patient revealing gross hyperostosis of the overlying bone; D) Clinical picture of the patient before the surgery after intubation revealing the 'mount meningioma'.

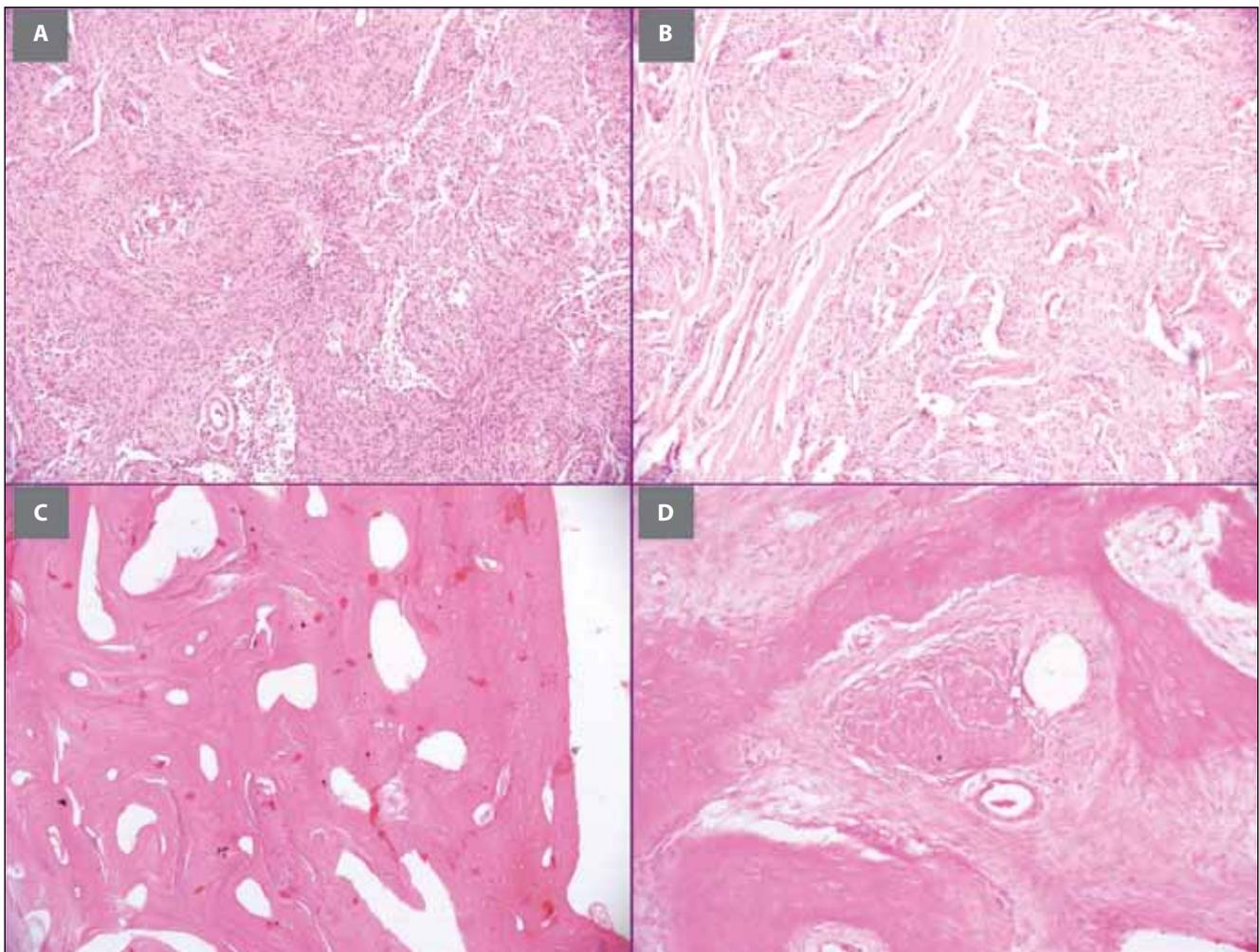


Figure 2: **A)** Photomicrograph from the intracranial tumor showing a transitional meningioma (HE, x100); **B)** Section from tissue over the hyperostotic bone shows similar tumor infiltrating the fibrocollagenous tissue (HE; x100); **C)** Section from hyperostotic bone shows markedly thickened bony trabeculae (HE; x100); **D)** At places, the tumor is seen infiltrating between the bony trabeculae (HE; x200).

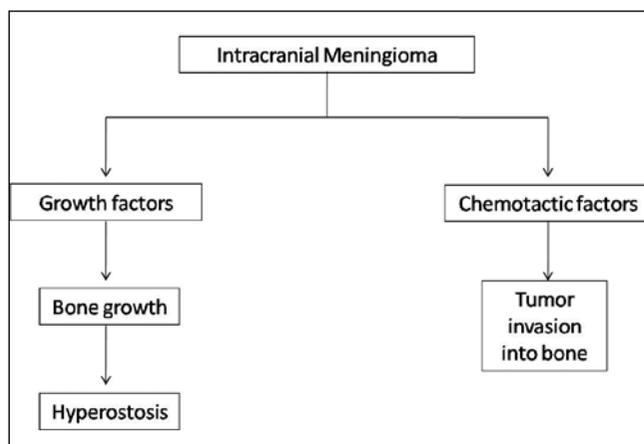


Figure 3: Common pathogenetic pathway leading to both radiological bone changes and tumor invasion into the bone.

DISCUSSION

Hyperostosis is known to occur in 25-75 % of meningiomas (5, 6, 9, 11, 16). The association between hyperostosis and meningioma was first described by Brissaud and Lereboullet in 1903 (3, 11). However, what causes hyperostosis is not known for sure. Various hypotheses have been proposed to explain the same including preceding trauma, tumor irritating the bone without invading into the bone, tumor cells secreting factors leading to stimulation of the osteoblasts, production of the bone by the tumor and vascular disturbances by tumor (15). An association between hyperostosis and tumor invasion was first suggested by Echlin in 1934 (8) and since then, tumor invasion has been postulated by many as the cause of hyperostosis (1, 2, 4, 7, 8, 10, 12-15).

In the present case, there was invasion of the overlying calvarium by the tumor cells with the tumor invading through the whole width of the bone and forming a *tumor cap* over the hyperostotic bone (Figure 1A-D). While it is known that the overlying bone is invaded by the tumor cells, it is still

not known whether tumor invasion is the cause of the bone changes or vice versa. It is possible that reactionary changes that occur in the overlying bone results in the production of growth factors, which induces bone formation, thereby leading to hyperostosis and chemotactic factors that attract the tumor cells into the bone matrix (Figure 3). Goyal et al. hypothesized that there might be common pathogenetic pathway, which results in both bone changes and tumor invasion into the bone (11). This case seems to support the above- mentioned hypothesis.

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