Childhood Malignant Meningioma

Çocukluk Çağında Malign Meninjioma

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Abstract: Childhood meningiomas, besides their rarity, grow fast, recur frequently and have a dismal prognosis when compared with adult tumors. A five year old patient presenting with headache, emesis and left hemiparesia was admitted to our hospital in 1990. On computerized tomography a 85x80x87 mm mass in the right frontoparietal region surrounded by significant edema, having a wide base on the falx causing 2 cm shift to left was found. The patient was operated on in our clinic with a diagnosis of multilobuled giant intracranial meningioma. The tumor showing frequent recurrence and sarcomatous changes is presented and discussed. Although the CT findings in this case suggest malignancy the behavior of the meningiomas is usually determined by histologic nature. In cases where frequent recurrence is seen the period between recurrences diminishes in time. Therefore CT controls are important in diagnosing recurrence. Total excision in early stage may increase the chance of survival.

Key words: Childhood, malignant, meningioma

Özet: Çocukluk çağı meninjiomaları nadir olmalarının yanısıra erişkin tümörleriyle karşılaştırıldığında hızlı büyüme, sık nüksetme, ve kötü bir prognoza sahip olma gibi özelliklere sahiptirler. Başağrısı, kusma ve sol hemipareziyle başvuran beş yaşında bir hasta 1990 yılında hastanemize yatırıldı. Bilgisayarlı tomografide sağ frontoparietal bölgede 85x80x87 mm boyutlarında belirgin ödemi olan, falksa geniş bir tabanla yapışmış, orta hatta 2 cm kaymaya neden olan bir kitle saptandı. Hasta, dev multilobule kafa içi meninjioma tanısıyla kliniğimizde ameliyat edildi. Sık nüks ve sarkomatöz değişiklikler gösteren tümör sunuldu ve tartışıldı. Bu olguda bilgisayarlı tomografi bulguları malignite düşündürmesine rağmen meninjiomaların davranışı genellikle histolojik yapısıyla belirlenir. Sık nüks gösteren olgularda nüksler arasındaki süre giderek azalır. Dolayısıyla aralıklı bilgisayarlı tomografi kontrolleri nüksü tanımada önemlidir. Erken dönemde tümörün tam çıkarılması yaşam şansını arttırabilir.

Anahtar sözcükler: Çocukluk, malign, meningioma

INTRODUCTION

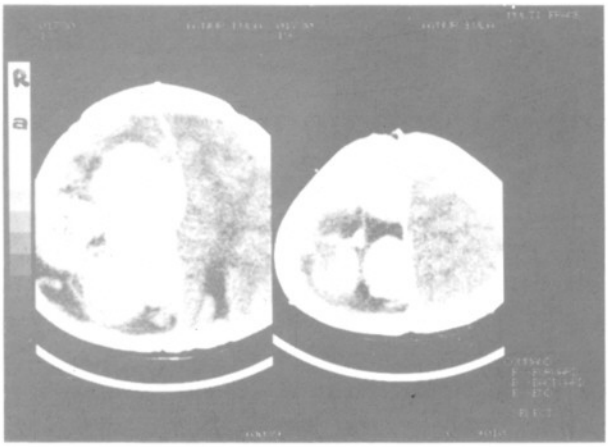
Intracranial meningiomas rarely occur in childhood and constitute about 0.4-3,5 % of all pediatric brain tumors (3,7,8,12,16,19,23,26). Matson reported only one patient in his series of 313 cases of intracranial tumors (8), and only three children with meningioma in his series of 750 cases of intracranial tumors under 14 years of age. In Cushing and Eisenhardt's series there are only six cases among 313 patients with intracranial meningiomas (3). Childhood meningioma series in the literature are shown in Table I (3,5,9,10,13,14,15,17,25,31).

We present a multilobulated giant intracranial meningioma case with frequent recurrence and sarcomatous changes.

CASE REPORT

A five year old girl complaining of headache, vomiting ,and left hemiparesis was admitted to our clinic in 1990. Her neurological examination showed bilateral papilledema, left sided spastic hemiparesis, increased deep tendon reflexes ,and Babinski sign. Preoperative computed tomography (CT) showed a mass situated in the left frontoparietal region, spontaneously hyperdense, intensely enhancing after contrast material injection, measuring 85x80x87 mm. The mass features fringes and irregular borders with a wide base on the falx (Figure 1,a and b).

The patient underwent a right central craniotomy and the mass was removed subtotally. Histological specimens revealed a meningioma with



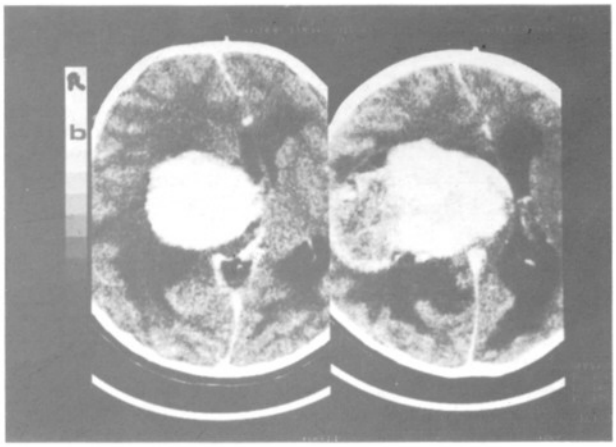


Figure 1, a and b: Preoperative computed tomography showed a multilobulated, hyperdense, intensely contrast enhancing, 85x80x87 mm left frontoparietal mass with fringes and irregular borders.

anaplastic features (Figure 2). Postoperative neurological examination did not show any additional neurological deficit. A CT scan performed

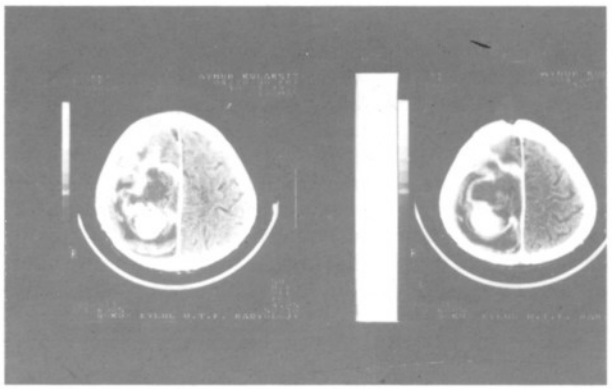


Figure 2: First postoperative contrast enhanced CT - showing residual mass and abscess formation.

17 days after the operation revealed residual mass and abscess formation (Figure 3). After antibiotic therapy for one month the abscess was drained and the culture revealed Staphylococcus epidermidis and aureus. Appropriate antibiotic therapy was given but a control CT two months later showed the mass and the persisting abscess. The patient was reoperated and the abscess was totally removed with residual mass. Histopatological examination revealed an anaplastic meningioma and secondary infection (Figure 4). Early and late control CT did not show any mass. Although radiotherapy was planned, the family refused further adjuvant therapy. Three months later the patient was readmitted to our clinic with severe headache. The CT showed a multilobulated heterogenous mass which was removed totally with repeat surgery. Postoperative course was uneventful and histopatological

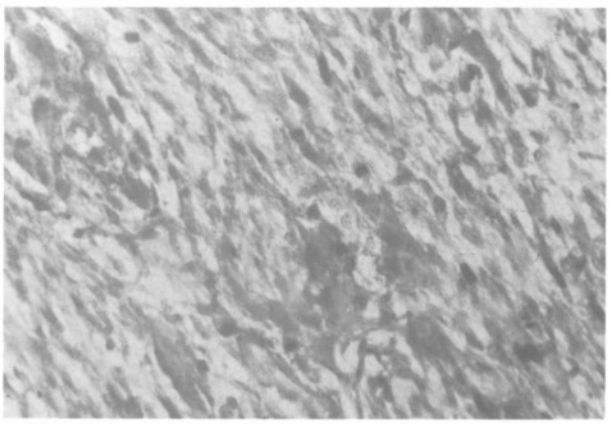


Figure 3: Histopathological specimens reveal an anaplastic meningioma. (H&E,X200)

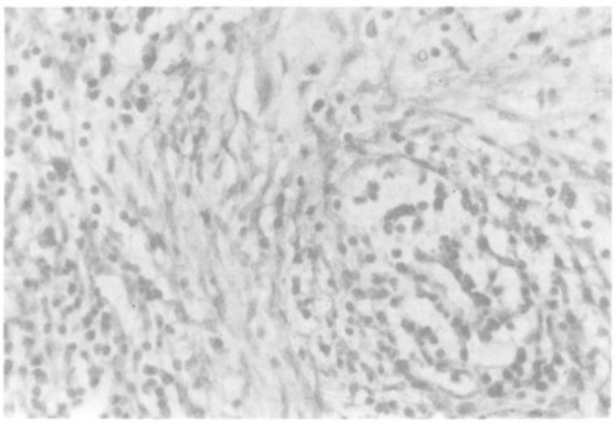


Figure 4: Histopathological examination reveals an anaplastic meningioma and secondary infection. (H&E,X200)

Table I. Childhood meningioma series.

Author	Age	Number of cases	Benign	Atypical	Malignant
Sosnik&Wrzesczynski (31)	At birth	1	1		
Fessard (14)	At birth	1	1		
Florin&Reid (15)	At birth	1	1		
Cuneo&Rand (9)	At birth	1	1		
Endo&Aihara (13)	3 days	1	1		
Benli at al. (5)	5 days	1	1		
Mendiratta at al. (25)	7 days	1	1		
Alp (3)	14 months	1		1	
Huang (17)	6 years	1		1	
Davidson (10)	4 months-16 years	27	23		4(2 sarcomatous)

examination showed meningioma with sarcomatous component. The control CT did not show any intracranial mass. Any further diagnostic study or therapy was refused by the family.

It was later learned that the patient had succumbed to the disease two years after the initial signs and symptoms.

DISCUSSION

Meningiomas are rarely seen in children (3,7,8,12,16,19,23,26) and differ from adult tumors by a tendency toward malignancy, increase in mass, and a worse prognosis (8,16,18). The incidence of intraventricular meningioma in children is reported to be higher than adults (17). Although the malignancy of meningioma is determined by histological examination CT may also hint at malignant behavior.

Computed tomography is an excellent method in the diagnosis of meningioma (4,16,27). Gd-DTPA enhanced magnetic resonance (MR) scans are considered to have a slightly higher diagnostic value than contrast enhanced CT (11,16,28,29). The typical CT findings are hyperdense or isodense mass with different rates of calcification, hyperostosis ,and edema showing contrast enhancement after contrast injection (4,11,16,19,27,28,29). The definite findings of malignancy and atypia are heterogeneous enhancement, hemorrhage, cyst formation, poorly defined or fringed margins, marked edema and osteolysis (4,11,16,19,29).

Computed tomography findings of this case are intratumoral hypodense areas, uncertain boundaries, fringes and extensive heterogeneous enhancement which seem to be the findings of malignancy.

Histopathological findings conclusively determine the malignant behavior of meningiomas (4,10). The extent of the surgical procedure (according to Simpson (1)) and the degree of anaplasia (presence of increased cellularity, loss of architecture, nuclear pleomorphism, mitotic figures, focal necrosis and brain infiltration) are the factors that effect tumor recurrence (1,4,19). The variants of meningioma in the new World Health Organization (WHO) classification (21) are meningothelial, fibrous, transitional, psammomatous and metaplastic (secretory, microcystic, clear cell, lymphoplasmacyte-rich) subgroups. The new classification includes atypical meningioma in the intermediate biologic behaviour group and in the malignant group, malignant and papillary types (21).

Malignant meningiomas retain enough histologic features to be recognized as meningiomas, but in addition have conspicuous mitoses, tumor necrosis, and invasion (21).

There are different grading schemes modifiying WHO criteria. In general grade I meningiomas are accepted as benign, grade II,III, and IV as atypical, anaplastic, and sarcomatous, respectively (6,22). The first and second biopsies of our case were evaluated as grade III. The last biopsy was evaluated as grade IV.

Anaplastic (malignant) meningioma can be recognized easily but may also be confused with anaplastic glioma, fibrosarcoma ,and schwannoma. Meningiomas are epithelial membrane antigen (EMA) and vimentin positive immunohistochemically and negative for glial fibrillary acidic protein (GFAP). Gliomas are positive for GFAP and fibrosarcomas are positive for vimentin but negative for EMA. Differention from schwannoma necessitate electron microscopy as well as positivity for S-100 protein (24). The tumor in our case was positive focally for EMA, and vimentin , but negative for GFAP and S-100 protein. Also meningothelial whorls, reticulin and collagen content were prominent focally in all of the biopsy materials.

After complete removal the 5 year recurrence rate is 78 % for anaplastic tumors (19) and again survival decreases in tumors with sarcomatous changes (1,4,19). Retrospective analysis showed that radiotherapy decreases recurrence rates and/or prevents recurrence (1,4,18). Metastatic meningioma is rare, constituting about 0.1 % of all meningiomas (2).

Radiation therapy was advocated in histologically malignant meningiomas (4,19). Again same retrospective studies showed a decrease in the recurrence rates of subtotally excised malignant meningiomas after radiotherapy (30).

Contrary to this idea, some authors believe that radiotherapy has some benefits, but it may have little value in the management of recurrent meningiomas (20).

Recurrence intervals get shorter every time, therefore it is important to follow the patient with periodic neuroimaging studies and to remove the tumor completely in order to prolong survival.

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