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MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY – RESEARCH PAPER WITH SYTAMATIC REVIEW

Neurosurgery

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ABSTRACT

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare pigmented neoplasm of infancy believed to originate from neural crest cells. These are rapidly growing locally invasive tumours which may occasionally show regional lymph node spread. Maxilla is the most common site of lesion followed by cranial vault and mandible. Treatment of choice for these tumours is surgical excision. Preoperative adjuvant chemotherapy can be helpful in patients with advanced disease. We report the successful surgical management of a six month old boy with a large frontoparietal lytic skull tumour which had extracalvarial and parenchymal extension

KEYWORDS

INTRODUCTION

Melanotic neuroectodermal tumor of infancy(MNTI) are rare pigmented osteoloytic tumours of infancy which can be surgically challenging. The invasiveness of the tumour, calvarial, extracalvarial and parenchymal involvement make surgical excision difficult. We report the successful surgical management of a 6-month baby boy with large tumour involving the frontoparietal bone at the anterior fontanelle. We also reviewed the literature concerning this type of tumor arising in the skull and discuss methods of treatment. 4,5

CASE REPORT

A 6-month-old boy was admitted with history of rapidly growing vault swelling since the last 2 months. There was no history of trauma or congenital anomaly. Examination revealed a we11-defined, nontender, nonpulsati1e, nonulcerated swelling at the anterior fontanelle. There was no palpab1e lymphadenopathy. No significant neurological findings were noted on examination. Laboratory tests were normal.Computed tomography (CT) and magnetic resonance imaging (MRI) showed an extracranial and intracranial lesion with involvement of the underlying dura mater. Metastatic workup was negative.

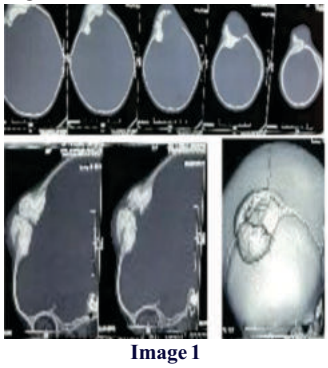

Image 1

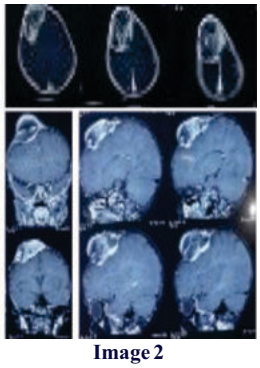

Image 2

Image 1 - Pre operative bone window of lesion involving right parental and frontal bone near coronal suture.
Image 2- Pre operative MRI images with contrast, enhancing lesion extension extra cranial and intra cranial compartment.

The baby boy underwent surgery under GA. The subcutaneous tumour was first dissected out, the infiltrated pigmented bone was then drilled until normal bone was exposed. The intracranial part of the tumor, which has invaded the underlying dura mater, was then totally excised. Intraoperativelt the tumor appeared blackish and has lobulated surface and infiltrating the underlying dura. There was a good dissection plane between the tumor and the brain. The entire tumor was excised and the boy had an uneventful recovery.

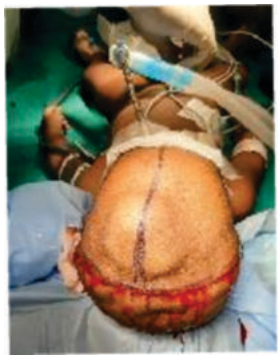

Image 3

Image 4

Image 3 - Surgical plan and marking of the incision.
Image 4 - Post operative follow up scalp site

Image 5

Image 5a

Image 5b

Image 5 - intra op lesion.
5a- intra cranial surface of the lesion with dura
5b - extra cranial surface of the lesion

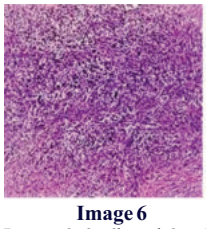

Image 6

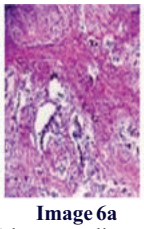

Image 6a

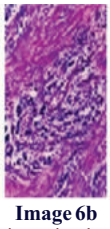

Image 6b

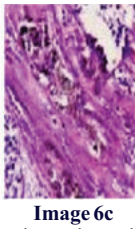

Image 6c

Image 6, 6a,6b and 6c - A hematoxylin eosin-stained specimen showed that the tumor was composed of a heterogeneous cell population, consisting of large epitheloid cells, mesenchymal cells and small rounded neuroblast-like cells. The mesenchymal component

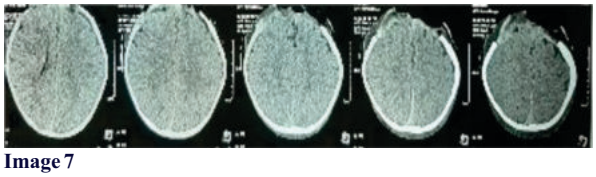

Image 7

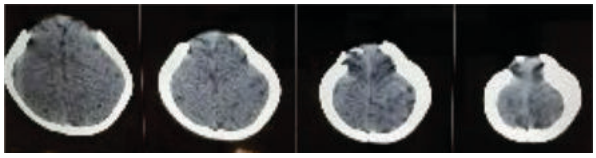

Image 7a
Image 7 - Immediate post operative image.
Image 7a- Follow up image after 6months

Histopathological examination revealed a melanocytic tumor. A hematoxylin eosin-stained specimen showed that the tumor was composed of a heterogeneous cell population, consisting of large epitheloid cells, mesenchymal cells and small rounded neuroblast-like cells. The mesenchymal component of the tumor cells had fusiform nuclei. The melanin-containing large epithelioid cells were diffuse positive with pan-CK and synaptophysin and focal positive with melen-A, but S-100 protein was negative.

DISCUSSION

Melanotic neuroectodermal tumor of infancy(MNTI) originate from neural crest cells and was first described by Krompecher E in 1918 as"melanotic progonoma," 1,4. MNTI is a rare osteolytic pigmented lesion of neural crest origin, predominantly seen in infants but may rarely be seen in older children and adults. The cell of origin is contentious and different theories have been proposed (Table). MNTI has a predilection for head and neck involvement 87%(2,3) with Maxilla (70%), skull(11%) and mandible (6%) accounting for majority of the cases. . It has also been documented in the extracranial sites such as the male (epididymus) and female reproductive organs, mediastinum, femur. In the skull, MNTI usually arises at and around the anterior fontanelle and near to the sutures. Brain parenchymal infiltration is usually secondary due to the extension of skull lesions, although cases of primary tumors arising in the cerebellar vermis and third ventricle have also been reported.

The MNTI clinically presents as a midline, painless, rapidly growing, pigmented, expansile, bony mass7,8. It primarily presents as a single lesion. However, multiple lesions have also been reported. Patients typically present with a nontender, nonulcerated, rapidly growing soft tissue swelling that results in skull deformation. The tumor causes compression rather than infiltration of adjacent structures.

Conventional radiographs of bony lesions usually show radiolucency with or without irregular margins. It is typical of CT scans to reveal hyper dense masses, but hypodense variants have been reported as well. The CT can accurately define the extent of the lesion and thus provides a good basis for surgical planning9,10. Magnetic resonance imaging shows a hypodense mass with focal areas of hyper-density9,10.

The cytology and histology are distinctive, showing a dual population of small neuroblastic cells and larger melanin-containing epithelial cells, as was seen in our case. IHC markers are helpful in differentiating MNTI from embryonalrhabdomyosarcoma (desmin and myoglobin positive), Burkitt's lymphoma (common leukocyte antigen positive) a n d malignant melanoma (HMB-45 and S-100 positive)11,12. Immunohistochemically, reactivity for HMB-45 and synaptophysin in our case indicate melanocytic and neuroblastic differentiationof the tumor cells, confirming the diagnosis ofMNTI 11,12.

The treatment of choice consists of complete surgical excision 13,14,15,16. Recurrence rates can be as high as 60%. . Most (540-70%) of recurrence happen from 4weeks to 4 months 3,6 and delayed resurgence of tumor is rare. Individuals with MNTI that are not amenable to surgical management alone may receive other modes of treatment 17,18,19,20,21. In genera1, this may be chemotherapy alone , chemotherapy with radiotherapy, chemotherapy before and after the surgical treatment, radiotherapy and surgical treatment or a combination of all. Chemotherapyay serve as an alternative or adjuvant option in the treatment of widely extended MNTIs.

CONCLUSION

Although MNTI behaves in a benign fashion, recurrences can occur especially within the first 6 months with the need for close follow-up postoperatively. Early detection and treatment will avoid further complications and may support a favorable outcome for the patient.In the present case, early diagnosis and treatment prevented further complications and the patient was followed-up for 24 months without any recurrence.

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