A CASE REPORT: MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY: DIAGNOSE AND TREATMENT

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ABSTRACT

Background: *Melanotic neuroectodermal tumour of infancy (MNTI) is a rare, pigmented neoplasm generally arising in infants during the first year of life. The tumor is rapidly growing, pigmented neoplasm of neural crest origin. The mean age of patients at diagnosis is 4.3 months, with a near-equal male to female ratio of 6:7.*

Material and method: *The aim of this work is to study one case of melanotic neuroectodermal tumour of infancy, has been diagnosed and treated at Pediatric center and the Odonto-Stomatology Center of Hue central hospital from 04/04/2018 đến 28/8/2018.*

Results: *A 8.5 month old male child presented with a mass in the anterior maxilla that had been growing since 2 months. The tumor grew rapidly. The child was done biopsy with the result showed melanotic neuroectodermal tumor. The child was removed the whole tumor, with the margin was negative. The patient was followed-up for 4.5 months without any recurrence.*

Conclusion: *Due to its rapid growth potential and locally destructive behaviour, early diagnosis is extremely important to limit local expansion. The treatment of choice for melanotic neuroectodermal tumor of infancy (MNTI) is surgical excision.*

Keywords*: Melanotic neuroectodermal tumor of infancy.*

I. INTRODUCTION

Melanotic neuroectodermal tumor of infancy (MNTI) is a very rare benign neoplasm arising in neonates and infants during the first year of life. A small number of cases have been reported in older children and adults. This tumor grows very fast and originates in the neural crest. MNTI generally originates from the soft tissue overlying the maxilla (68-80%), but it can occasionally arise in the skull (10.8%), mandible (5.8%) or brain (4.3%). In addition to the head and neck region, other sites can be affected by the condition less frequently, including the femur, epididymis, ovaries, uterus and mediastinum. Clinically, MNTI is soft and reddish-blue tumor. It often destroys the underlying bone and prevents teeth development. Clinical and radiological findings can suggest a diagnosis of MNTI, but pathology is golden criteria. The mean age of patients at diagnosis is 4.3 month, with a near equal male to female ratio of 6/7. The best choice for treatment is surgical excision. The rate of recurrence ranges from 10-15%, and malignancy rate of 6.5% is reported in the literature [1], [3].

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II. PATIENT AND METHOD

2.1. Patient

A 8.5 month male child presented with a mass in the anterior maxilla that had been growing since 2 months. He was admitted hospital when he was 4 months. He was diagnosed melanotic neuroectodermal tumor of infancy.

2.2. Method

Describe one case

III. RESULTS

3.1. Clinical and subclinical presentations

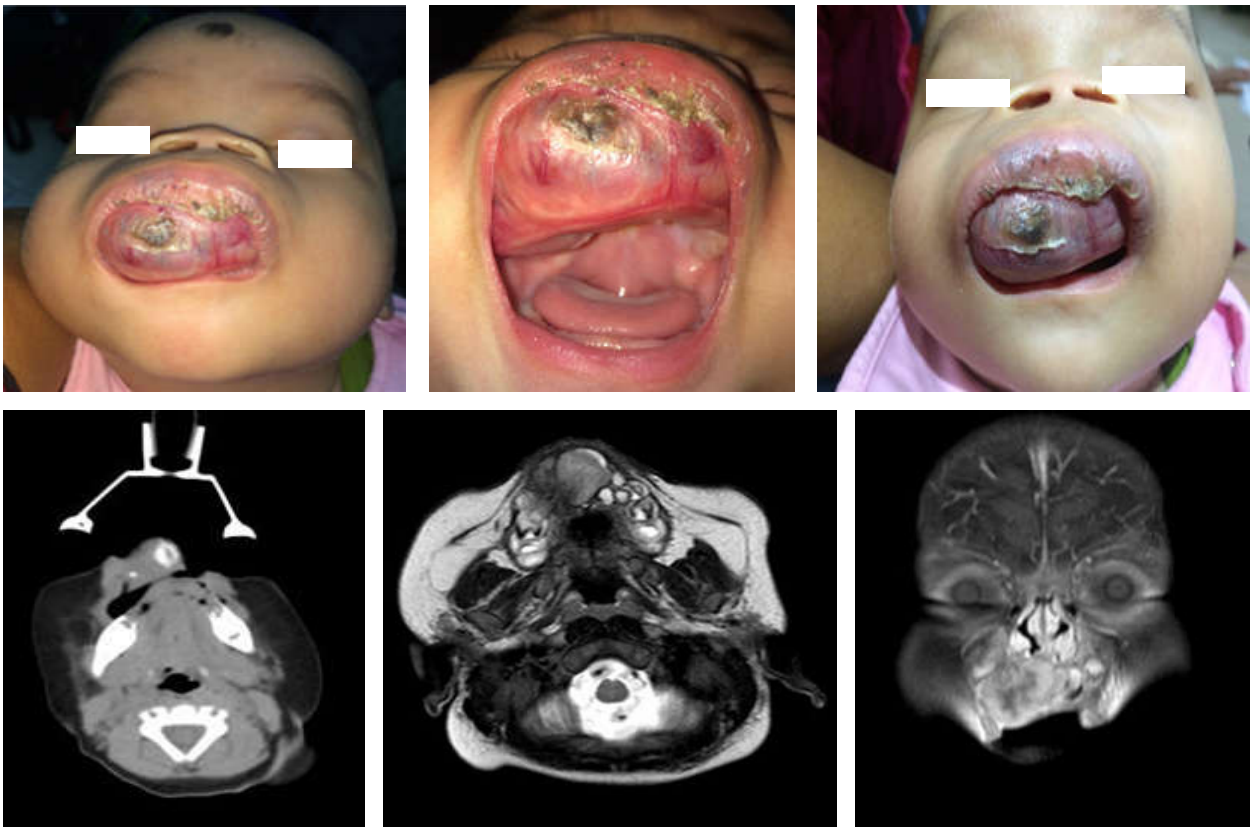
Clinical presentation: Patient H, male, was born on 17th December, 2018 at Phu Vang district, Hue city. The child was admitted Hue Pediatric Center on April, 2018 due to the tumor at his maxilla.

The symptoms at the admitted hospital time:

He was alert

Facial asymmetry with the tumor in the anterior maxilla and hard palate. The tumor was oval, non-ulcerative, no pain, reddish-blue tumor. The tumor was protruded through the lip, with the dimension was about: 2x3x4 cm.

The patient can not suck, his mother had to put small milk spoon to his mouth. Other organs were normal.



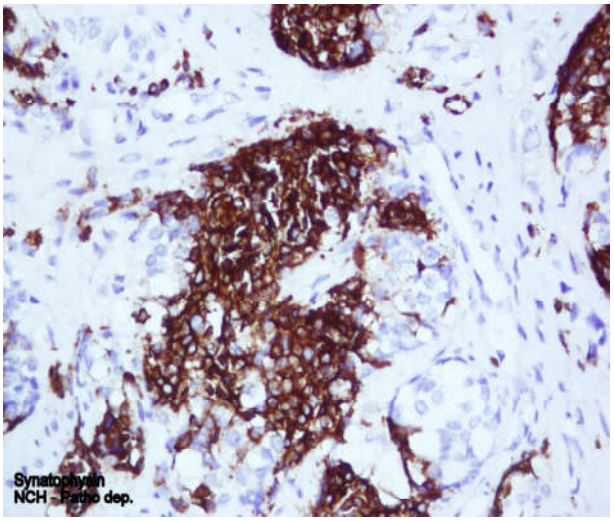
MRI result:

The MRI scans showed one heterogenous lesion at the right anterior maxilla with dimension: 22x28x35mm. The lesion showed hyperintense signals on T2W images. The lesion destroyed maxilla and hard palate.

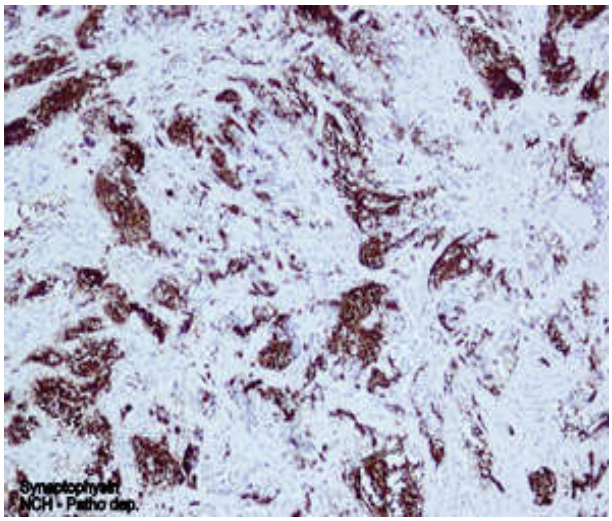
The result of CT 64 of whole body: There is not any metastasis.

Pathology result:

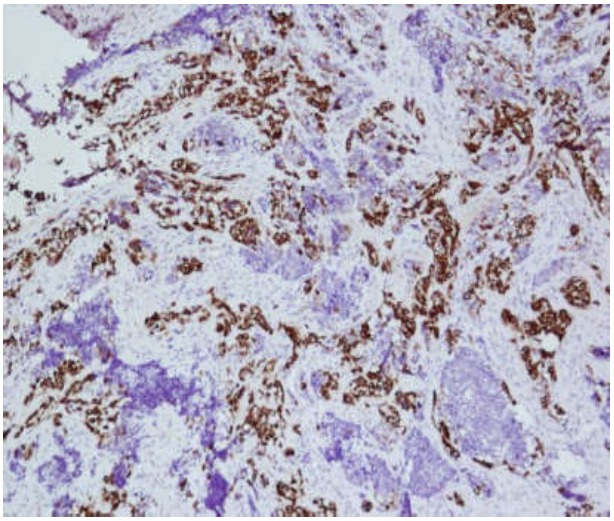
At the beginning, we did needle biopsy, pathologists in Hue Central Hospital read it, then we sent to National Hospital to review. After surgery, we sent the sample to St Jude Children Hospital, Memphis, USA to review. The results showed the proliferation of a dual population of cells: 1/ polygonal cells wich have pigment and they are positive for HMB45, EMA; 2/ small round cells groups with neurofilament background, they are positive for Synaptophysin and CD56.



Synaptophysin (+)



CD 56 (+)



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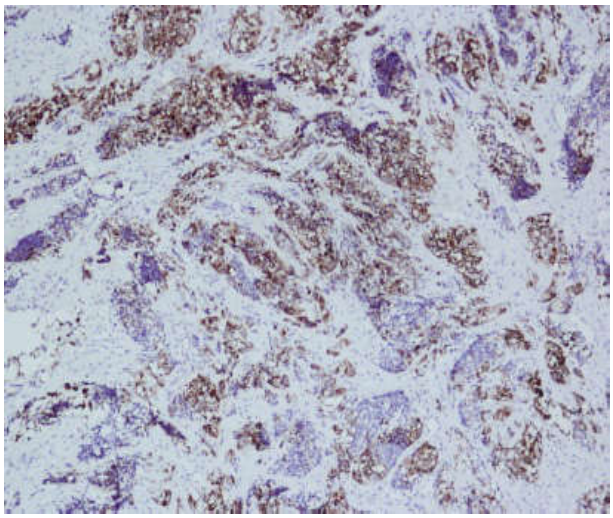


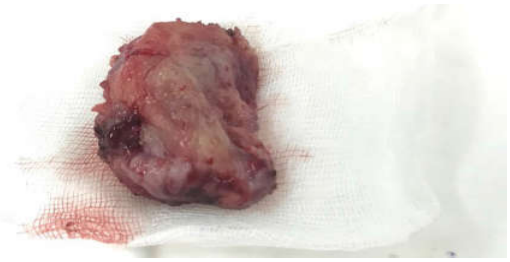
HMB 45 (+)

Diagnosis: Melanotic neuroectodermal tumor of infancy

3.2. Treatment

The baby was discussed with doctors from St Jude Children's Research Hospital and we decided to remove the tumor completely. This was the first time we met this case, we didn't have experience. So, we contacted with Prof McKay McKinnon from Chicago Hospital, USA. On 17th May 2018, Prof McKinnon came to my hospital and did surgery for the baby with some doctors from the Odonto-Stomatology Center. The tumor was removed totally.

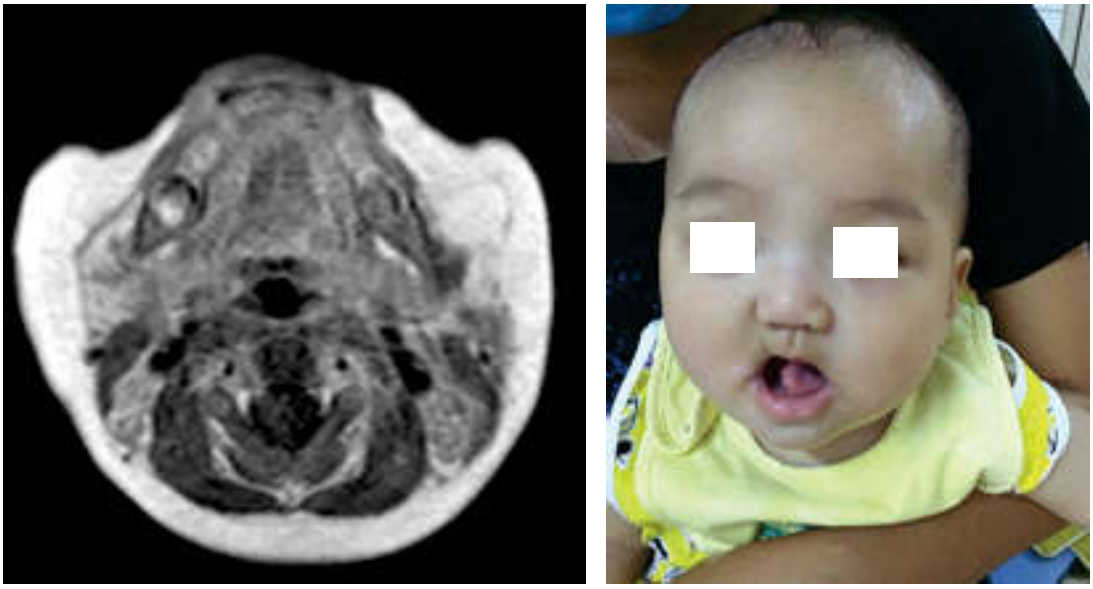
The report of operation:



The pictures after surgery

The patient was taken under general anaesthesia. An incision was made in the mucosa along the boundaries of the tumor. The tumor was resected totally with one part of maxilla and peripheral excision with 2 mm margin. The margin was done cold biopsy with the result was negative.

After surgery, the baby recovered soon. Two months later, he was checked with MRI. The result showed no residual tumor. At now, 4.5 months after surgery, there isn't any sign of recurrence.



The pictures after surgery

IV. DISCUSSION

4.1. Diagnosis

MNTI generally occurs in the craniofacial region with a predominance for the maxilla (68-80%), followed by the skull (10.8%), mandible (5.8%). Other sites can be affected, such as: brain (4.3%), epididymis, mediastinum, ovary, uterus and peripheral bone. The MNTI is never congenital. It emerges within the 1st year of life, mostly below 6 months of age. Infants present with a painless, non-ulcerative bluish black gingival mass that is often confused with an eruption cyst. It may appear to be malignant due to its rapid growth potential [2], [4].

Our patient manifested tumor within the first two months of birth, and he was brought to our hospital when he was 4 month. The tumor located at the anterior maxilla, with characteristics: non-ulcerative, no pain, reddish-blue tumor. And this was the first time, we met this tumor. We didn't have any experience, so we thought the tumor could be lymphoma, or rhabdomyosarcoma or tooth enamel tumor. After having the pathology result, we knew this disease and we reviewed this case again and we found out that the patient had symptoms suited MNTI.

At the time patient admitted hospital, we did some tests and tomography and magnetic resonance imaging (MRI) scans. Then we did needle biopsy. The MRI scans showed one heterogenous lesion at the right anterior maxilla with dimension: 22x28x35mm. The lesion showed hyperintense signals on T2W images. The lesion destroyed maxilla. And the biopsy result showed two phases of tumour cells: 1/ polygonal cells wich have pigment and they are positive for HMB45, EMA; 2/ small round cells groups with neurofilament background, they are positive for Synaptophysin and CD56. With these results, a definitive diagnosis of MNTI was made.

To assess metastatic, the result of whole CT scan showed that the patient didn't have any signs of organ metastatic. Some researches showed metastatic is rare, occuring in only 3% of patients. When metastases develop, the smaller neuroblatic cels predominate in the secondary deposits. Histology therefore resembles neuroblastoma more than MNTI [5]

4.2. Treatment

MNTI has a significant destructive nature with high growth potential and are furthermore unencapsulated. In this patient, the tumor was first

noticed when he was 2 month old. He was admitted hospital when he was 4 months. The tumor was growing very fast, especially at the time he was in hospital. We saw the tumor grew and changed its dimension every week. The treatment of choice for MNTI is surgical excision, and it is usually curative. Existing teeth and developing teeth must be sacrificed when they lie within the lesion or near the borders of an MNT1. Some controversy still exits regarding the amount of adjacent bone that needs to be removed during the surgical procedure. Some clinicians approved that peripheral excision with 2-5 mm margin were generally considered as ideal. Others advocate for only enucleation of the tumor followed by curettage of the bone cavity. However, a recent systematic review claims that no differences in recurrence rates were observed between curettage and resection [5]. In this patient, when doing surgery, we chosed the tumor removal with 2 mm margin from the tumor. The margin was done cold biopsy with the result was negative.

4.3. Prognosis

At 5 years after diagnosis, both sexes showed equal relapse rates. The overall incidence of local recurrence was 10-15%. Currently, age at manifestation is a considered to be strong prognostic indicator in MNTI. Infants who manifested within the first two months of birth were associated with a high risk of recurrence which generally occurred within 6 months from treatment. In contrast, manifestation from 2.5 to 4 months was associated with an intermediate risk and manifestation after 4.5 months of age had a minimal risk of recurrence [6], [7]. And most of recurrence cases could be saved by extensive resection. Chemotherapy is not a usual treatment procedure except for cases of confirmed metastatic diffusion.

Regarding our patient, the baby manifested within the first two months of birth, so he belongs to high recurrence group. And now, he hasn't had any recurrence symptoms since 4.5 months after surgery. However, we need to follow up him carefully to find out and treat any early recurrence [8].

V. CONCLUSION

Melanotic neuroectodermal tumor of infancy is usually a benign tumor arising from cells of neuroectodermal origin. Due to its rapid growth potential and locally destructive behavior, early diagnosis is extremely important to limit local expansion and the extent of required tissue resection. However, the rarity of the tumor often leads to a delay in diagnosis, resulting in a less than desired outcome.

This was the first case MNTI we met at Hue Central Hospital. With the good cooperation between some centers inside, as well as the collaboration with oversea experts that helped us to have prompt diagnosis and good treatment for the patient. The patient will be followed up in one year to assess any recurrence.

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