ISSN: 0976-3031

Available Online at http://www.recentscientific.com

CODEN: IJRSFP (USA)

International Journal of Recent Scientific Research
Vol. 9, Issue, 1(L), pp. 23718-23720, January, 2018

International Journal of Recent Scientific Research

DOI: 10.24327/IJRSR

CASE REPORT

DIAGNOSTIC DILEMMA: MELANOTIC NEUROECTODERMAL TUMOUR OF INFANCY A CASE REPORT

Pallavi S and Saini Mondal*

Department of Oral and Maxillofacial Pathology

DOI: http://dx.doi.org/10.24327/ijrsr.2018.0901.1523

ARTICLE INFO

Article History:

Received 16th October, 2017
Received in revised form 25th November, 2017
Accepted 23rd December, 2017
Published online 28th January, 2018

Key Words:

Melanotic, tumour, immunohistochemistry and recurrence.

ABSTRACT

Melanotic neuroectodermal tumour of infancy (MNTI) is a rare, pigmented rapidly growing neoplasm of neural crest origin. This tumour primarily affects the maxilla and surgical excision is considered as the treatment of choice. Here we report a case of MNTI in a 6month old baby girl with histopathological features, immunohistochemical features and a brief review. A 1 year follow up was done and no recurrence was noted.

INTRODUCTION

Melanotic neuroectodermal tumour of infancy (MNTI) is a rare and benign melanin-containing mesenchymal tumour of neural crest origin with higher recurrence rate.[1,2,3] MNTI is usually evident within the first year of life with no gender predilection.[4]Clinically it is seen as rapidly growing mass with partial pigmentation and expansile nature.[5] MNTI have higher prevelance for anterior maxilla.[1,2,3,6,7,8] Various term has been used to describe this lesion like congenital melanocarcinoma, retinal anlage tumour, pigmented congenital epulis or melanotic progonoma.[1,5,9] MNTI was first described by Krompecher in the year 1918 and later in the year 1966, it was suggested that the tumour had neural crest cells origin due to excretion of large amounts of vanillylmandelic acid (VMA) by Borello and Gorlin.[3,7,9,10,11,12] The histological analysis of the MNTI shows close resemblance of the cells to that of neuroblasts and electron microscopic studies shows presence of neurosecretory granules which together classify this tumour to be of neural crest origin.[10]

Although histopathologic examination is considered reliable for the final diagnosis of MNTI but is not enough to differentiate benign and malignant variant as cellular atypia along with sparse mitosis can also be noted in the benign counterpart.[3]Histopathological hallmarks considered for diagnosis of MNTI are epithelioid and neuroblastoma-like biphasic differentiation of tumor cells and existence of melanin pigment.[13] The immunohistochemical analysis can be performed for the final confirmation of the difficult cases.[3,9,10] The immunohistochemical markers which shows positivity are cytokeratin, HMB45, where the pale stained cells exhibits melanocytic differentiation and synaptophysin , NSE where the small darkly stained cells exhibit neuroblast like differentiation. It also shows positive results for epithelial membrane antigen, glial fibrillary acidicprotein and Leu-7, but shows negativity for S-100 protein.[3,10].

Here we present a case with brief review of Melanotic Neuroectodermal Tumor of Infancy.

CASE REPORT

A 6month old baby girl presented with a swelling in the maxilla. The swelling was initially small and gradually grew to attain the current size over a period of 1month.The swelling started obstructing the oral cavity resulting in difficulty of feeding but was painless(Fig 1). No other relevant medical history was found. Growth and development of the infant was normal for her age. Clinical examination revealed a well defined dome shaped, bluish tinged swelling on the right side of the anterior gum pad region size 2x1.5 cm extending antero-posteriorly from labial vestibule to mid-palatal region. No

Corresponding author:* **Saini Mondal
Department of Oral and Maxillofacial Pathology

evidence of ulcerations, bleeding, pus discharge or sinus opening was found.

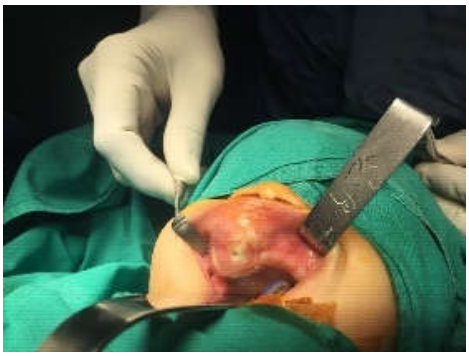

Figure 1 Clinical Image

Radiographically a hazy radiopacity of decreased density was noted in the pre- maxilla along with labial expansion (Fig 2).

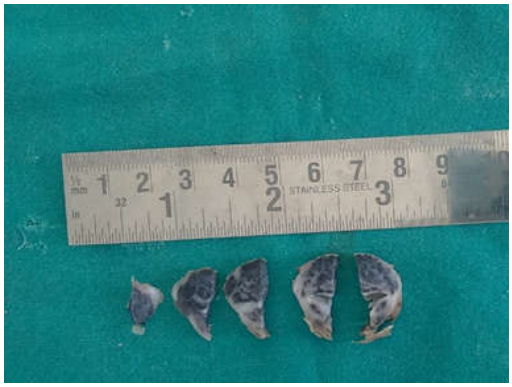

Figure 2 Gross Image of MNTI

A provisional diagnosis of eruption cyst and vascular neoplasm was given. An excisional biopsy was done and gross examination showed firm, well circumscribed mass measuring about 2x1.5 cm and cut surface of the specimen was grayish black in colour(Fig 3).

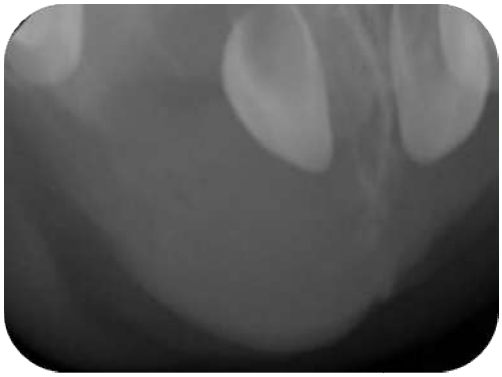

Figure 3 Radiographic Image

Microscopic examination(H&E) showed non encapsulated mass composed of dual population of small round blue cells and larger melanin containing epithelial cells within the dense fibro cellularstroma(Fig 4) Epithelial cells were larger with vesicular nuclei and abundant cytoplasm containing melanin granules, arranged in alveolar and tubular pattern with few of them encircling the small round cells that were predominantly arranged in nests and sheets with small hyperchromatic nucleus and scanty to fibrillary cytoplasm.

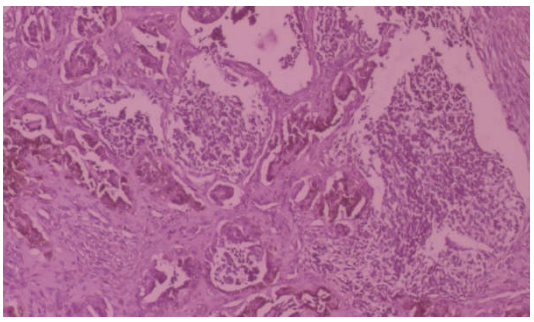

Figure 4 H/E in 10x showing dual cell population of small round cells and larger melanin containing epithelial cells within the dense fibrocellular stroma.

The tumour was seen entrapping bony trabeculae and very scanty mitotic activity was also noted.

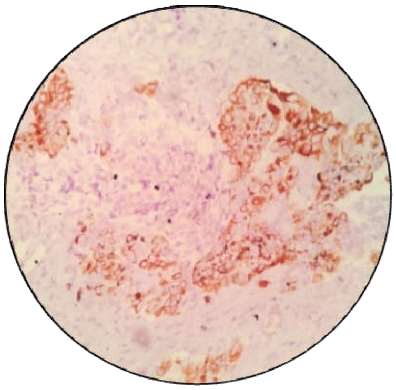

Figure 5 10X showing CK positive for melanocytic cells

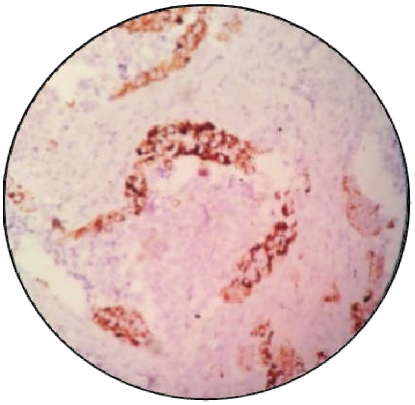

Figure 6 10X showing HMB45 positive for peripheral melanotic cells

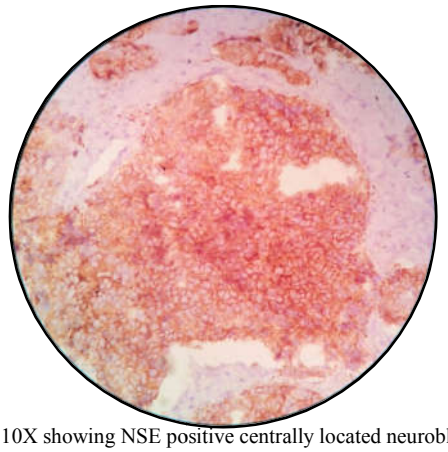

Figure 7 10X showing NSE positive centrally located neuroblast cells

Immunohistochemical study revealed positive staining for CK (cytokeratin) (Fig 5), HMB-45(human melanoma black 45) (Fig 6) and NSE (neuron specific enolase) (Fig 7) markers. Both the neuroblast cells & epithelial cells expressed NSE strongly while only the epithelial cells expressed HMB-45 (cytoplasmic) and CK (both membranous & cytoplasmic) positivity. Neuroblast cells were strongly negative for CK.

DISCUSSION

Melanotic neuroectodermal tumour of Infancy(MNTI) was first described by Krompecher in the year 1918 when it was termed as congenital melanocarcinoma.[1,4,6,8] Later it was renamed as Melanotic Neuroectodermal Tumour of Infancy by Borello and Gorlin which is universally accepted till date.[6] Occurrence of MNTI is seen predominantly in first year of life with equal gender predilection.[4,7] A rapidly growing non-ulcerated mass in cranio-facial region affecting mostly anterior maxilla being the primary site is noted in most of the cases.[3,5,6,8] The tumour causes melanin synthesis but it might not be clinically evident.[8] Several theories have been proposed for the explanation of pathogenesis of this neoplasm that resembles early stages of retinal development that is like that of retinal anlage tumour.[11] Other studies have shown neural crest cell origin for this lesion where the neural cells are seen differentiating into cell types like neuroblast and melanocytes.[5,10] Other tumours of neural crest origin that resembles retinal an lage tumour are pheochromocytoma, neuroblastoma and ganglioneuroblastoma.[11] Inspite of the aggressive behaviour of the MNTI it is still considered as benign tumour, though some studies considered it to be potentially malignant due to the recurrence which is about 20% and metastasis rates around 3-7% about in those studies.[3,5,6,8] The differential diagnosis of MNTI includes the developmental cysts like the nasopalatine cyst and globulomaxillary cyst.[3,5,10] Odontogenic lesions like the odontoma, ameloblastoma, ameloblastic fibroma, odontogenic myxoma, adenoameloblastoma and odontogenic keratocyst are also considered. The non-odontogenic non-neoplastic lesions includes central giant cell granuloma, fibrous dysplasia and arteriovenous malformation. Non-odontogenic neoplastic lesions like rhabdomyosarcoma, Burkitt lymphoma, Langerhans cell histiocytosis and Ewing sarcoma are also considered as differential diagnosis.[3,5,10]Histopathologically it shows biphasic nature where small round cells and epitheloid cells containing melanin pigments arranged in nest like or alveolar pattern.[4,5,6,8] These features were consistent with our case where epithelial cells were larger with vesicular nuclei & abundant cytoplasm containing melanin granules that were arranged in alveolar and tubular pattern. Immunohistochemical studies shows positivity for HMB45 which stains the melanin containing epitheloid cells and synaptophysin positivity also which stains the neuroblast like cells. [1] These features of immunohistochemical study was again similar our case. MNTI stain spositive for Cytokeratin which was also similar to our case[9]

The treatment modality is complete surgical excision with minimum of 5mm healthy margins.[1,2,3] Chemotherapy has also been used for the treatment purpose but for general consideration it can be considered only in cases where patients are not amenable for the surgical excision.[1,13] Delayed recurrences has been observed in many of the cases so a follow up is always advised for the cases of MNTI.[1]

References

1. Agarwal P, Saxena S, Kumar S, Gupta R. Melanotic neuroectodermal tumor of infancy: Presentation of a case affecting the maxilla. *Journal of oral and maxillofacial pathology*: JOMFP. 2010 Jan; 14(1):29.
2. Bangi BB, Tejasvi MA. Melanotic neuroectodermal tumor of infancy: A rare case report with differential diagnosis and review of the literature. Contemporary clinical dentistry. 2012 Jan; 3(1):108.
3. Nazir B, Gupta H, Chaturvedi AK, Rao SA, Jena A. Melanotic neuroectodermal tumor of infancy: Discussion of a case and a review of the imaging findings. Cancer Imaging. 2009; 9(1):121.
4. Chaudhary A, Wakhlu A, Mittal N, Misra S, Mehrotra D, Wakhlu AK. Melanotic neuroectodermal tumor of infancy: 2 decades of clinical experience with 18 patients. *Journal of Oral and Maxillofacial Surgery*. 2009 Jan 1; 67(1):47-51.
5. Tan O, Atik B, Ugras S. Melanotic neuroectodermal tumor in a newborn. *International journal of pediatric otorhinolaryngology*. 2005 Oct 1; 69(10):1441-4.
6. Choudhury M, Mukherjee J, Bajaj P, Jain A. Melanotic neuroectodermal tumor of infancy. Indian pediatrics. 1997 Mar 1; 34:248-50.
7. Anil S, Raji MS, Beeana VT. Melanotic neuroectodermal tumor of infancy. Report of a case. *Indian journal of dental research: official publication of Indian Society for Dental Research*. 1993;4(2):65-7.
8. Hamilton S, MacRae D, Agrawal S, Matic D. Melanotic neuroectodermal tumour of infancy. *Canadian Journal of Plastic Surgery*. 2008 Mar;16(1):41-4.
9. Andrade NN, Mathai PC, Sahu V, Aggarwal N, Andrade T. Melanotic neuroectodermal tumour of infancy–A rare entity. *Journal of oral biology and craniofacial research*. 2016 Sep 1;6(3):237-40.
10. Pattanayak S, Ray JG, Mukherjee S, Mandal C, Chaudhuri K. Melanotic neuroectodermal tumour of infancy. BMJ case reports. 2010 Jan 1; 2010: bcr0120102645.
11. Shakeel M, Shafi M, Imran M, Ahad M. Melanotic Neuroectodermal Tumor of Infancy. *Oral & Maxillofacial Pathology Journal*. 2016 Jan 1; 7(1):694-7.
12. De Oliveira MG, Thompson LD, Chaves AC, Rados PV, da Silva Lauxen I, Sant'Ana Filho M. Management of melanotic neuroectodermal tumor of infancy. *Annals of diagnostic pathology*. 2004 Aug 1; 8(4):207-12.

How to cite this article:

Pallavi S and Saini Mondal.2018, Diagnostic Dilemma: Melanotic Neuroectodermal Tumour of Infancy A Case Report. *Int J Recent Sci Res.* 9(1), pp. 23718-23720. DOI: http://dx.doi.org/10.24327/ijrsr.2018.0901.1523

****