

# Melanocytic Neuroectodermal Tumour of Infancy of Testis - A rare Case Report

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## ABSTRACT

**Introduction:** Melanocytic neuroectodermal tumor of infancy (MNTI) is an uncommon pigmented neoplasm of neural crest origin, occurs mainly in the head and neck region specially maxilla. It shows slight male predominance. Melanocytic neuroectodermal tumor (MNTI) of testis is rare, proper diagnosis and differentiation from small round cells tumours (SRCT) is important as treatment modalities of these tumors are quite different.

**Case report:** Here we present a case of MNTI of testis in an eleven month old male which was clinically diagnosed as teratoma. A distinctive morphology and immunohistochemistry play important role in postoperative diagnosis of this tumor.

**Conclusion:** Treatment of MNTI is surgical excision with clear margins and follow up.

**Keywords:** Neuroectodermal, Pigmented, Testis, Tumour

## INTRODUCTION

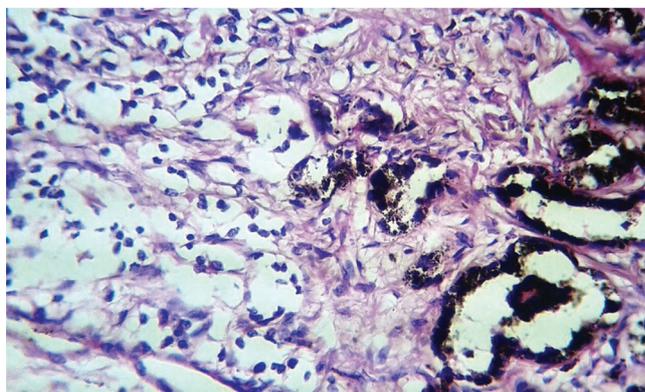
Melanocytic neuroectodermal tumour of Infancy in the testis is first described by Krompecher in 1918 as congenital melanocarcinoma.<sup>1</sup> Currently, based on biochemical ultrastructural and immunohistochemical studies it is thought to be a neoplasm of neural crest derivation.<sup>2</sup> Majority arises in head and neck region (in over 90%) mainly in maxilla but may arise in other sites.<sup>3</sup> Melanotic neuroectodermal tuomr of infancy(MNTI) arising in testis is extremely rare.<sup>4</sup> Maximum number of cases developed in the first year of life and both sexes are affected equally.<sup>5</sup> Clinical courses is generally benign but local recurrence rate is 10-15% and metastasis have been documented in less than 5% cases.<sup>3</sup> Surgery remains the mainstay of treatment for patient with MNTI.<sup>6</sup> We report a case of MNTI arising in left testis with complete resection (left high inguinal orchidectomy) in a 11 month old boy which was clinically and radiologically suspected as teratoma. Later on postoperative histopathological and immunohistobhemical study confirmed the diagnosis of MNTI.

## CASE REPORT

An 11 month old boy was referred to the department of Paediatric Surgery, Gauhati Medical College and Hospital (GMCH) for evaluation of slow growing scrotal mass which was initially noticed by parents. Birth and family histories were unremarkable. Clinical examination showed normally developed male infant (weighting 8kg). Local Examination revealed single fixed non tender well circumscribed left testicular lesion/mass approximatedly 3.5X3 cm sized. There was no inguinal lymphadenopathy. The child was admitted in the paediatric surgery department



**Figure-1:** Gross Specimen of testis with testicular mass showing dark brown pigmented area centrolaterally



**Figure-2:** Small cells forming alveoli admixed with pigmented large cells forming glandular structures (H&E X400)

for evaluation. Ultrasonographic study (USG) showed well defined echogenic lesion measuring 3.5X3 cm in the left testis. All other laboratory test were within normal limit. Xray chest, CT abdomen and pelvis were within normal limit without any evidence of metastasis. Based on clinical and radiological finding a provisional diagnosis of teratoma was made and subsequently the child underwent left high inguinal orchidectomy with complete excision of the mass.

Gross pathologic examination of the testicular mass revealed 3.5x3.5x3 cm<sup>3</sup> sized specimen (Fig-1), round relatively

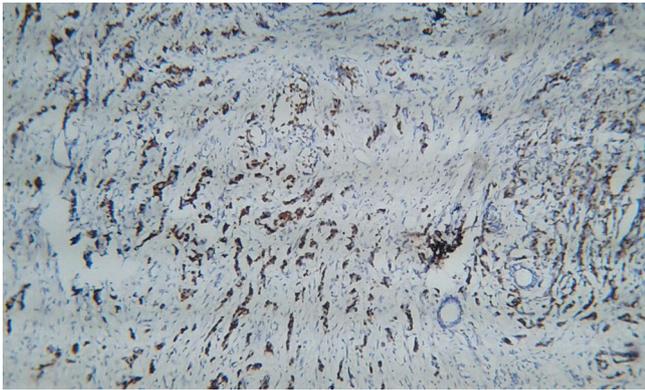
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**Figure-3:** Immunohistochemical stain (IHC) showing Chromogranin positivity in small cells (IHC X100)

well circumscribed whitish tumor mass. Cut surface is whitish with patchy ill defined dark areas of pigmentation involving the testis. Microscopical examination showed the tumor was composed of biphasic population of cells, small round neuroblastic cells (nested and alveolar pattern) and large pigmented epithelioid cells forming tubules or pseudoglandular pattern at places. Large cells have abundant cytoplasm enlarged nuclei with prominent nucleoli. Some of them contain intracytoplasmic pigments. Small neuroblast like cells have scanty cytoplasm with small dark nuclei (Fig. no. 2). Mitosis were rare and there was no extension of the tumor cells to the nearby tissue and it had pushing border. Immunohistochemically small cells were positive for chromogranin (Fig. no.3) and epithelial cells were positive for cytokeratin. On basis of histopathology and IHC diagnosis of MNTI arising in testis was made. Postoperative period was uneventful, patient was discharged and regular follow up was advised. Till date patient is doing well without any evidence of recurrence or metastasis.

## DISCUSSION

Melanotic neuroectodermal tumor of infancy (MNTI) is an uncommon pigmented neoplasm of neural crest origin, 92.8% occur in the head and neck region with nearly 70% arising within the the maxilla.<sup>7</sup> It was first described by Krompecher in 1918 as melanocarcinoma. Since then there have been numerous change in its nomenclature because of uncertainty regarding its origin or histogenesis. The term melanotic neuroectodermal tumor of Infancy was first used by Borello and Gorlin in 1962 with elevated level vinyl mandalic acid (VMA).<sup>8</sup> Misugi et al and Nikai et al further supported the neural crest theory based on the ultrastructural morphology.<sup>9,10</sup>

The first description of MNTI occurring in epididymis of testis was reported by Eaton and Ferguson in 1956.<sup>11</sup> Melanocytic neuroectodermal tumor of infancy (MNTI) usually present as firm lobulated well circumscribed mass compressing but not infiltrating the adjacent structures. Size range from few centimetre to 13 cm in diameter. The cut surfaces usually white containing dark central area.<sup>1</sup> Histologically two population of cells are there, small round cells and pigments containing large cells.<sup>2</sup> Because of its rarity chances of misdiagnosis is common. Important

differential diagnosis are small blue round cells tumors like neuroblastoma, rhabdomyosarcoma, ewings sarcoma, peripheral neuroectodermal tumor (PNET), lymphoma and malignant melanoma. A distinctive morphology and demonstration of the various epithelial, melanocytic and neural markers aid in the diagnosis of MNTI.<sup>2</sup> Increased expression of Ki67 and CD99 has been correlated with more aggressive behaviour.

Surgery remains the treatment of choice.<sup>6</sup> Recurrence has been known to occur in 10 -20% of cases.<sup>3</sup> Therefore surgical excision with clear margins is mandatory. Occasionally metastasis was described in regional lymphnode, liver pleura, bone marrow, ureter, urinary bladder, pelvis, bowel. Postoperative chemotherapy and radiotherapy were administered to patients with MNTI with metastasis.<sup>1</sup> In our case there was no any evidence of regional and distant metastasis and simple high inguinal orchidectomy was done.

## CONCLUSION

Melanocytic neuroectodermal tumor of infancy (MNTI) usually follows a benign course and most cases are treated with simple excision. Since treatment modality and prognosis of MNTI and small round cell tumor (SRCT) are very different, it is important to make correct diagnosis. Here histopathology and immunohistochemical studies plays vital role and support the theory of neural crest origin.

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