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GIANT CONGENITAL NEVUS WITH PLEXIFORM NEUROFIBROMA AND EPITHELOID VARIANT OF MALIGNANT PERIPHERAL NERVE SHEATH TUMOR WITH MELANOCYTIC DIFFERENTIATION A RARE CASE REPORT

RANJANA GIRI, SUBRAT SAHU, K.L PUROHIT, J RATH &U SENAPATI

Department of Pathology & Surgery, KIMS, Campus-6, Patia, BBSR, Orissa, India

ABSTRACT

A 22 year male patient with a giant congenital nevus in association with plexiform neurofibroma and epitheloid malignant peripheral nerve sheath tumor (MPNST) on the trunk and back. This is a very rare entity, till now only two cases has been reported in the literature. The clinical, surgical and histological aspects of the disease are discussed. The mutual origin of all the three entities can be explained by boland theory of neurochristopathy.

KEYWORDS: Giant Congenital Nevus, Plexiform neurofibroma, Epitheloid MPNST

INTRODUCTION

A male patient with a giant congenital nevus associated with plexiform neurofibroma and malignant peripheral nerve sheath tumor is presented. The giant congenital nevus is a benign tumor and a variant of congenital melanocytic nevus characterised by its extensive size measuring more than 20cm in its greatest dimension and occupying >20% of body surface area, often there are many scattered satellite lesions associated with them. It may also have inclusion of other cells of the neural sheath e.g Schwann cells, ganglion cells and pleomorphic nevoid cells[1]. Patients who present with LCMN are known to have high risk of developing malignant melanoma[2,3,] rhabdomyosarcoma, malignant peripheral nerve sheath tumor,lipoma and neurofibroma. The concept of neurocrystopathy by bolard has proven that maldevelopment of the neural sheath differentiated during the embryologic phase of evolution may lead to varying combination of tumor diseases after parturition.[1]

Neurofibroma are defined as a benign tumors of the nerves sheath which along with Schwann cells, perineural cells and fibroblast cells and it may have inclusion of different cells that may cause the problem in defining the origin of the cell population in a malignant alteration[2]. Plexiform neurofibroma is a variant of neurofibroma showing nodular architecture with myxomatous change. Plexiform neurofibroma is very rare but has the greatest potential of malignant alternation among all benign peripheral nerves tumors ranging from 5-10%[4].Only two case having combination of the giant congenital nerves, plexiform neurofibroma and MPNST has been reported till date.

MATERIALS & METHODS

22 year old male presented with congenital giant nevus in the body with multiple satellite pigmented lesions in both the hands and also in both legs (Figure 1). The patient began developing numerous small rubbery nodules over the nevus since childhood which was slowing growing in nature. Since last 6 months there is sudden enlargement of two nodules occure for which became for medical attention. (Figure 2). Biopsy was taken from the two large nodules and sent for histopathological study. First specimen was a skin covered tissue of size 3x2x2cm. Cut section yellowish lobulated,

firm, homogenous and at places glistening (Figure 3). Second specimen was a irregular oval tissue of size 2x1.5x1.5cm. Cut section brownish black and soft in consistency (Figure 4).

RESULTS

Section from the first lesion shows a tumor with nodular architecture seen in the dermis. Nodules are of verying size(Figure 5) with conspicuous myxoid stroma with stellate and oval shaped tumor cells (Figure 6). Few small aggregates of melanin containing cells are also seen. There is no pleomorphism or mitosis seen.Immunohistochemistry was diffuse and strong positivity for S100, HMB-45 is positive In pigment containing cells and desmin negative, Histopathological diagnosis was plexiform neurofibroma.

Section from the second lesion shows a tumor arranged in sheets, nests with epithelioid cytomorphology (Figure 7). Tumor cells are round to polygonal with distinct cytoplasmic border some showing prominent nucleoli.few mitosis seen. At places the tumor cells are seen with perivascular orientation (Figure 8). Few melanin containing cells are also seen (Figure 9). IHC shows S100 diffuse and strong positivity. HMB-45 was positive only in pigment containing cells. EMA and pancytokeratin were negative.

Correlating the clinical history, histological finding of the other lesion as plexiform neurofibroma the histopathological features of the second lesion is considered as epitheloid MPNST with melanocytic differentiation. One year after biopsy the patient is well.

DISCUSSION

Giant congenital nevus with plexiform neurofibroma and MPNST is a very rare entity it mostly occurs during childhood and adolescence[4,5]. The origin of this entity can be studied in two different stand points. Malignant peripheral nerve sheath tumor, nevus and plexiform neurofibroma from one entity in embryologic and pathogenetic sense and according to bolande it could be defined as a complex form of neuro cristopathy and neuricristopathic syndrome, the subgroup being sundry association and interrelated complex[1]. Which can be explained on the basis of a defect in the neural crest which is considered to be a common origin of melanoblasts, Schwann cells, sensory ganglia, bone, fat, muscle and blood vessels. When associated with neurofibromatosis, the 5- years survival rate is 16% when isolated the 5 year survival rate is 53%.our case shows a rare form of epithelioid MNPNT and plexiform neurofibroma a over a large congenital nevus.only two similar case in the literature was described, one by drazen shejbal et al on 2012[6]. They described an 15 years female with giant congenital nevus associated with plexiform neurofibroma and malignant peripheral nerve sheath tumor. Another case by Roth et al who described an infant girl born with a pigmented giant nevus developed a malignant schwannoma in the retroperitoneum at 16 months of age.[7]

CONCLUSIONS

The index case is very rare and unusual in respect to its clinical presentation. Till date only two cases has been reported. But Giant congenital nevus with plexiform neurofibroma & Epitheloid MPNST with melanocytic differentiation is not reported till now. *Of all benign* tumors of the peripheral nerve origin, plexiform neurofibromas with the risk of malignant alteration greater than 5%, represents the tumor with the highest malignant potential. Our case shows a rare form of plexiform MPNST in a patient without neurofibromatosis. Only histopathology & immunohis to chemistry are overlapping between malignant melanoma and Epitheloid MPNST. corelating the clinical history, his to morphology of

first lesion as plexiform neurofibroma we diagnosed the case as epitheloid MPNST with melanocytic differentiation. The giant nevus and MPNST are physically connected with transitional areas of the plexiform neurofibroma confirming the common origin of all three tumors.

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APPENDICES

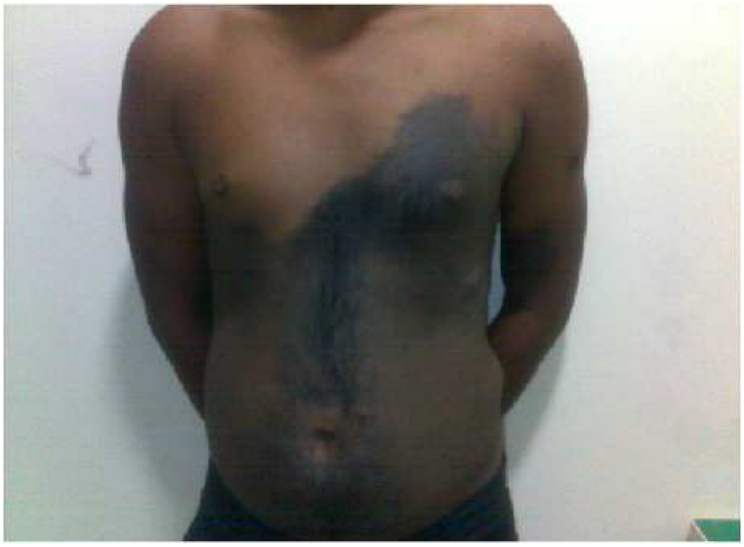

Figure 1

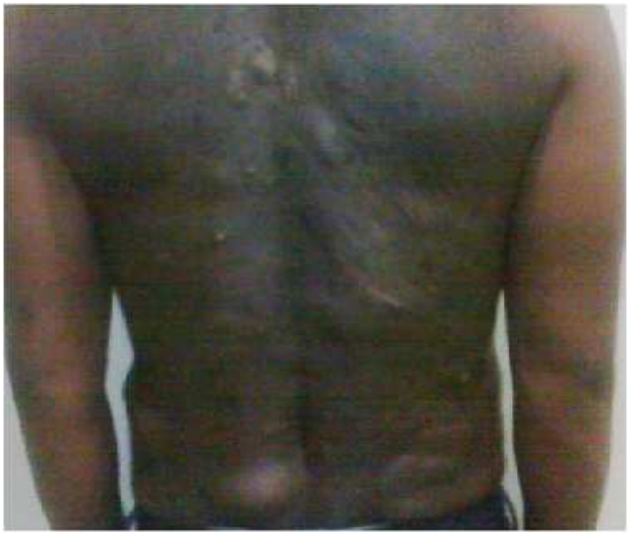

Figure 2

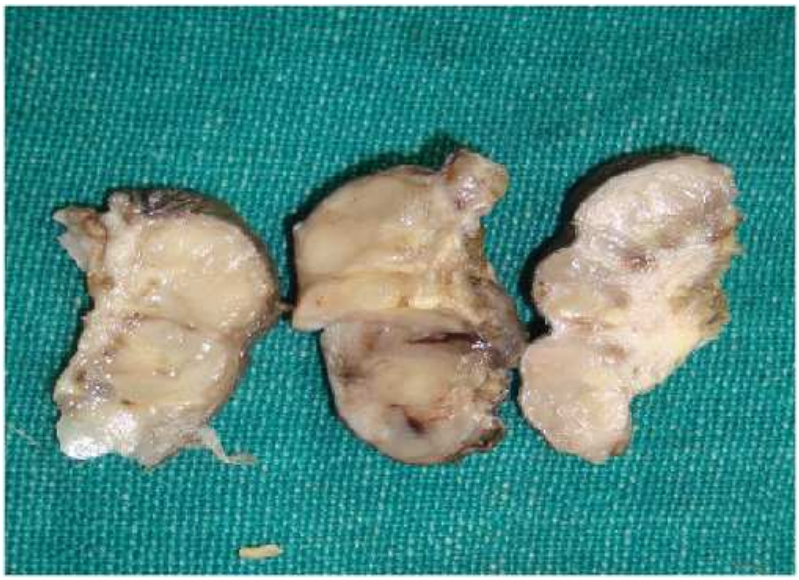

Figure 3

Figure 4

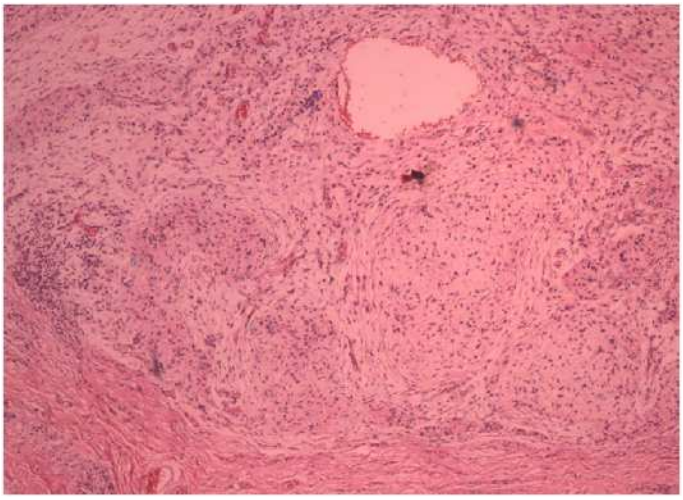

Figure 5

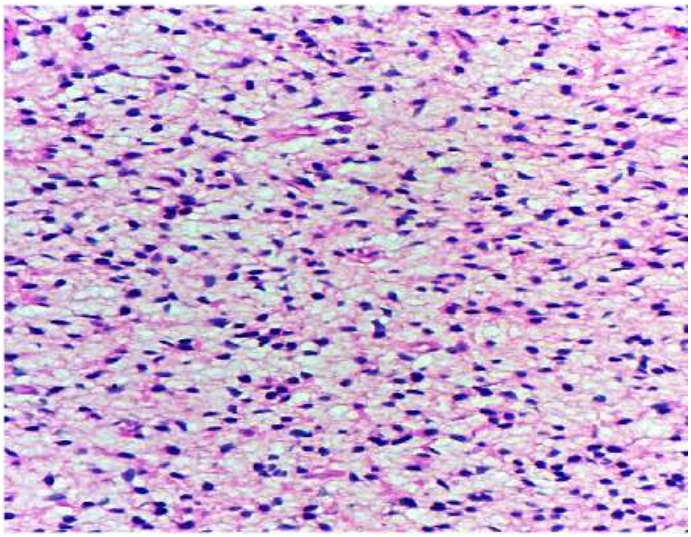

Figure 6

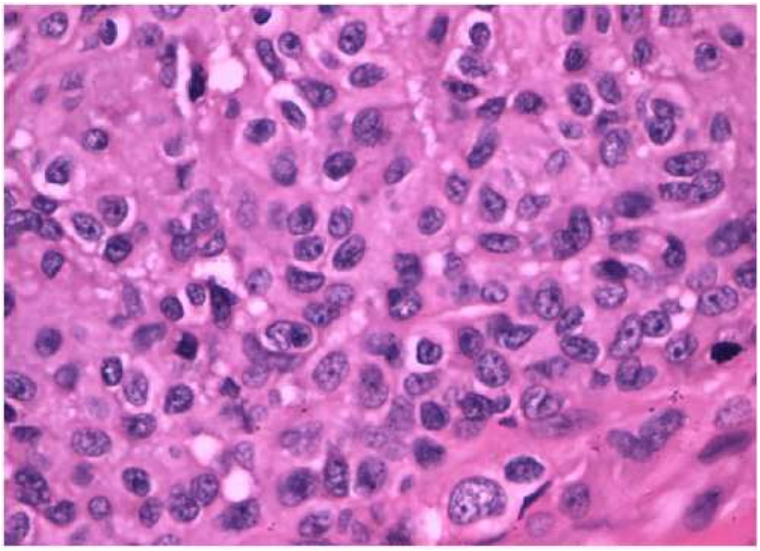

Figure 7

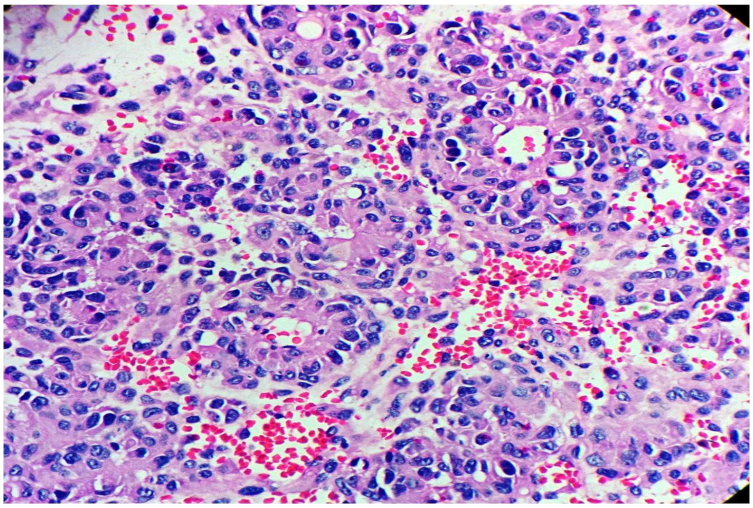

Figure 8

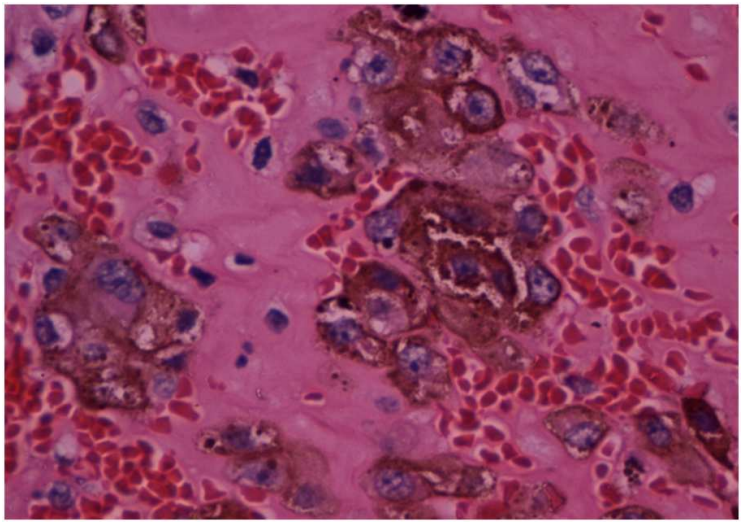

Figure 9