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PENILE NEUROFIBROMA IN A 10-YEAR - OLD CHILD: A RARE CASE STUDY

Mohammed Jaseem Hassan, Saqib Ahmed

Department of Pathology

Jawaharlal Nehru Medical College, Aligarh Muslim University, U.P., India-202002

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ABSTRACT

Neurofibromas of the penis occur uncommonly and their solitary occurrence is even rarer. We report a case of a ten year old child presenting with a solitary neurofibroma located on penis. A provisional clinical diagnosis of penile retention cyst was made and histopathological examination releaved a benign spindle cell lesion and few close differential diagnosis were made, which on application of immunohistochemistry was diagnosed as neurofibroma of penis. Surgical excision of the lesion was done and there was no recurrence after a year of follow up.

Address for correspondence

Dr. Saqib Ahmed
Department of Pathology
Jawaharlal Nehru Medical College,
Aligarh Muslim University, India-202002.
Email: Saqibahmed.mail@gmail.com
Contact no: +91-9411800648

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INTRODUCTION

Neurofibroma is a benign peripheral nerve sheath tumour which consists of perineural cells, schwann cells, fibroblasts and mast cells in a myxoid background and arises from endoneurium and the peripheral nerve sheath (1). Mutation in the NF1 gene has been found as the aetiological root of neurofibroma (2). Neurofibromas are mainly of three types and include the localized type (most common), diffuse type and the plexiform type,each having a very low malignant potential except the plexiform type which carries an increased risk of malignant transformation and is usually associated with neurofibromatosis type 1 (NF 1) (2). Neurofibroma of the penis, especially the solitary form is very rare and is usually associated with neurofibromatosis type 1 (3). To our knowledge, till date about tewnty one patients with primary neurofibromas associated with penile neurofibromatosis have been reported, with its limited occurrence in children (4). To improve the understanding, diagnosis and treatment of the disease, we report a case of penile neurofibroma.

CASE REPORT

A 10-year-old male presented with a complaint of swelling at the penile tip for two months with a slow progressive enlargement. He had no previous history of trauma. Physical examination revealed no abnormality in the abdomen and/or genitourinary system examination. No associated lymph node was palpable.No family history of similar swelling was revealed. A provisional clinical diagnosis of penile retention cyst was made and an excision biopsy of the cyst was done and was sent to the Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh. On Gross examination, a single grey-brown soft tissue piece measuring 1x0.5x0.3cm was received. Entire tissue was submitted in a single block for further histopathological processing. On histopathological examination of the section fascicles of ovoid to spindle cell tumour with spindle-shaped plump nuclei to wavy nuclei with clear to eosinophilic cytoplasm were seen (Fig. 1.1,1.2)The tumour cells were also arranged concentrically around blood vessels with a myxoid background (Fig. 1.3). A broad diagnosis of Benign Spindle cell Lesion was made and a differential diagnosis of Neuroma/neurofibroma, Fibromatosis, Myopericytoma and Myofibroma were kept. Immunohistochemistry for Smooth Muscle Antigen (SMA), S-100, Beta Catenin was done for exact categorisation.SMA was negative (Fig. 2.1) S-100 showed patchy positivity in spindle cells (Fig. 2.2) while Beta-catenin was negative for nuclear staining (Fig. 2.3). Thus a final diagnosis of the Neurofibroma of the penis was made.No recurrence was reported after a follow up of one year.

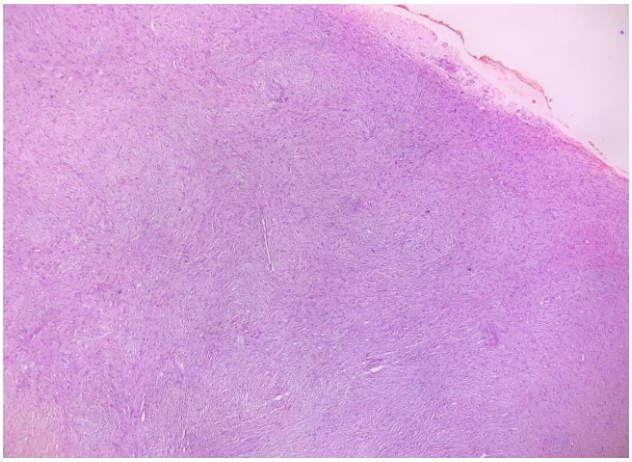

Fig.1.1: Section Shows Well Circumscribed Proliferating Bundles of Interlacing Spindle Cells (H & E X10)

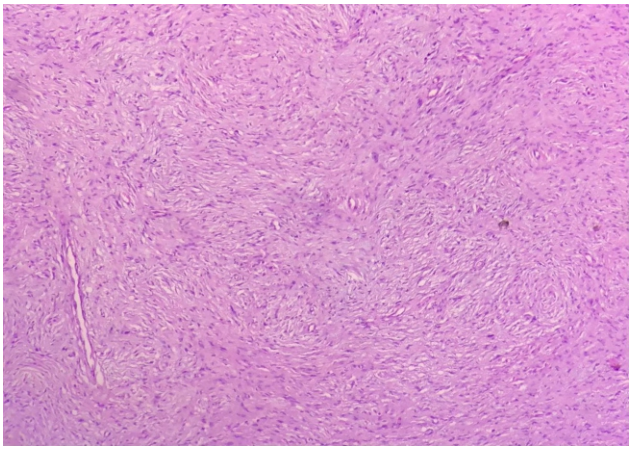

Fig.1.2: Section Shows Spindle Cells in Interlacing Bundles with Interspersed Collagen (H and E X10)

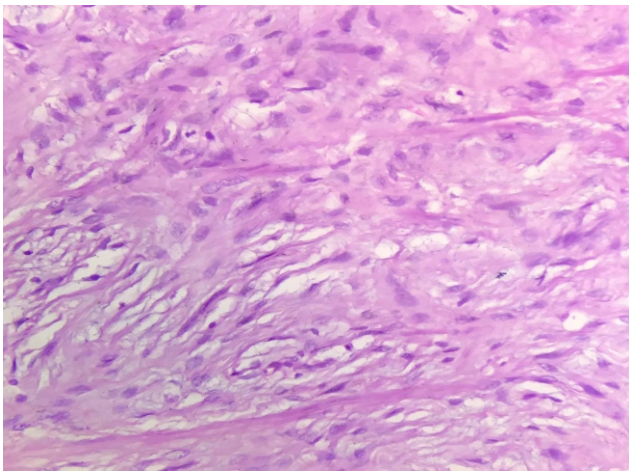

Fig. 1.3: Section Shows Ovoid to Spindle Cells with Wavy Nuclei with Clear to Eosinophilic Cytoplasm Arranged Concentrically around Blood Vessels with a Myxoid Background

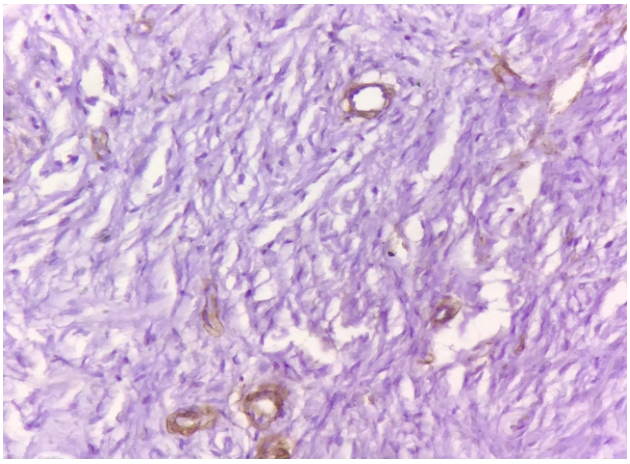

Fig 2.1: Section Shows Negative Staining for Smooth Muscle Antigen (SMA) Immunostain (X40)

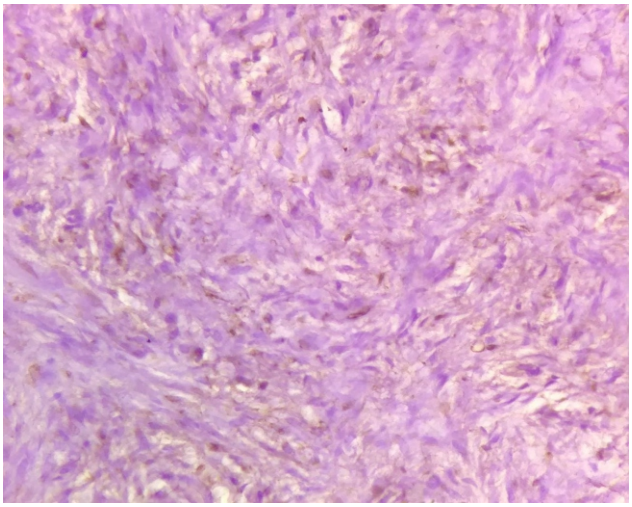

Fig. 2.2: Section Shows Patchy Positive Staining in Spindle Cells for S-100 Immunostain (X40)

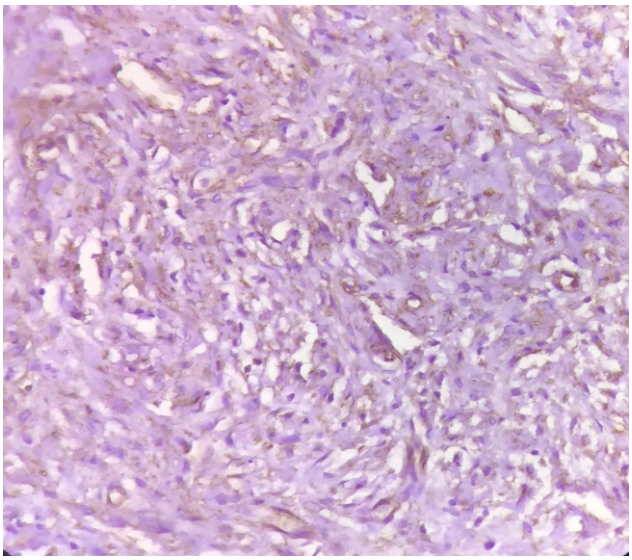

Fig. 2.3: Section Shows Negative Nuclear Staining for Beta Catenin Immunostain (SMA) (X40)

DISCUSSION

Neurofibroma is a neural tumour, benign in origin that arises from the peripheral nerve sheath and is composed of perineurial-like cells,schwann cells and fibroblasts in variable amounts (5). They are usually found in the young population and may be either solitary or multiple depending on the number of neurofibromas in the body (6).Most Neurofibromas are have no association with neurofibromatosis type 1 and termed as solitary, while multiple neurofibromas are usually associated with neurofibromatosis (6). Neurofibromas usually occur in the head and trunk region with few cases involving the colon, bladder, spermatic cord, stomach and other extremities (7) Neurofibroma of the penis is quite rare and usually show association with neurofibromatosis type 1 (8). Histopathological differentials on similar locations like penile fibromatosis, Myopericytoma and Myofibroma should be carefully ruled out to avoid misdiagnosis. Penile Fibromatosis is characterised by dense fibrous nodules of myofibroblasts along with hyalinizing fibrosis and perivascular lymphoid infiltrates and is usually positive for vimentin, beta-catenin and variably positive for SMA but negative for S100 (9). Myopericytoma is a spindle cell neoplasm of low grade potential and shows ovoid shaped tumour cells and moderate to abundant eosinophilic cytoplasm with perivascular proliferation. Immunohistochemically it is positive for SMA but negative for S-100 (10). Myofibroma is a well-circumscribed, nodular neoplasm characterized by a distinctive biphasic growth pattern in which the centre of the lesion is composed of immature appearing plump spindle-shaped tumour cells along with hemangiopericytoma-like branching blood vessels while the periphery is shown nodules and fascicles of variably hyalinized and myoid appearing cells and show positivity for SMA and negativity for S 100 (11). Macroscopically neurofibroma is usually well circumscibed, unencapsulated, grey to tan appearance,firm with a pale gelatinous cut surface (12). While microscopic examination features includes spindle-shaped cells arranged in an interlacing pattern with round to fusiform nuclei and moderate to abundant eosinophilic cytoplasm within a loose matrix of fibrillary collagen and a myxoid background (13). Localised type of neurofibroma is usually well-circumscribed lesion (14), while the diffuse type is poorly defined and shows expansile proliferation with characteristic pseudomeissnerian corpuscles which are composed of fibrillary and whorled Schwann cells (15). Plexiform type of neurofibroma shows nerve fascicles showing hypertrophy with thick collegen fibres in a predominantly myxoid to edematous background (16). Immunohistochemical features of Neurofibroma shows S100 positivity in Schwann cells (approximately 50% of tumour cells) while CD34 positivity in spindled fibroblasts (usually greater than 60% of the tumour cells).EMA may be positive in occasional perineural cells. The treatment of NF is mainly surgical resection, and the prognosis is usually good.

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Orcid ID:
Saqib Ahmed - http://orchiod.org/0000-0003-0545-1735
Mohammed Jaseem Hassan - http://orchiod.org/0000-0002-7256-7175

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