

Case Report

Rare site and size of a neurofibroma: a case report

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ABSTRACT

Neurofibromas are tumor of neural connective tissue, mainly composed of Schwann cells and fibroblasts. They can occur anywhere in the body, presenting as a feature of neurofibromatosis or as a solitary mass except in the brain and spinal cord. A large neurofibroma in the inguino-scrotal region is a rare condition with few reports available in the literature. We report a case of a 32 year old male with a large mass extending from the inguinal region into the thigh, mimicking an indirect inguinal hernia. Since he had multiple nodules on his body suggestive of neurofibromatosis, a diagnosis of benign neurofibroma was made, the mass excised and the diagnosis confirmed by histo-pathological examination. Elongated, spindle shaped neoplastic cells with poorly defined cytoplasm was observed. No evidence of atypical mitotic figures noted.

Keywords: Inguinal, Scrotal, Neurofibroma

INTRODUCTION

Neurofibromatosis is a tumor of neural connective tissue, arising from large and small nerves, presenting an autosomal dominant pattern of inheritance. Available literature suggest multiple criteria for the diagnosis of neurofibroma, including presence of Café au lait spots, freckling in the axillary and inguinal region, multiple nodules of neurofibromas occurring anywhere in the body causing severe disfigurement and discomfort.^{1,2} New mutations occur in about 50% of patients with neurofibroma, making these gene loci one of the highest known mutation sites in humans. Neural tissues are found throughout the body, these tumors can occur in a variety of sites.³ Although it can be encountered anywhere within the central or peripheral nervous system, especially in the neck, thorax, cranium, retroperitoneum, and flexor surfaces of the extremities, localization within the scrotum is extremely rare.⁴ Present case reports a large neurofibroma of the anterior abdominal wall extending down the scrotal region into the thigh, mimicking an

indirect inguinal hernia, causing extreme discomfort for the patient while standing as well as sitting, with added fungal infection of the skin over the swelling and the thighs.

CASE REPORT

A 32 year old male presented with a swelling in the right groin, of 3 years duration. It was pain less, gradually increasing in size, extending down the scrotal region into the thigh, causing extreme discomfort while standing and sitting. General examination of the patient revealed multiple, small nodules all over the body, suggestive of familial neurofibromatosis. Local examination revealed a large mass measuring 10 cm in breadth X 20 cm in length descending from the right inguinal region, literally displacing the scrotum and testis to the left (Figure 1). It was pedunculated, firm, non-tender, not attached to the scrotal skin or other underlying structures. Transillumination negative. Inguinal nodes not palpable. Fungal infection was present on the skin over the

swelling and also on the medial side of the right thigh. Routine hematological and biochemical investigations were within normal limits. Because of the extreme discomfort suffered by the patient, a total excision of the pedunculated inguino-scrotal mass was done under spinal anesthesia. Wound healed well and the post-operative period was uneventful. He was discharged on the third post-operative day and advised periodic follow-up. Histopathological examination confirmed the diagnosis, showing elongated, spindle shaped neoplastic cells with poorly defined cytoplasm was observed (Figure 2). No evidence of atypical mitotic figures noted. Consent of the subject was taken as per human ethical guidelines from the Apollo Hospitals, Ranipet, Tamil Nadu. (AH/RP/TN/21/8/2014).



Figure 1: Large mass of neurofibroma extending into the scrotal region mimicking indirect inguinal hernia.

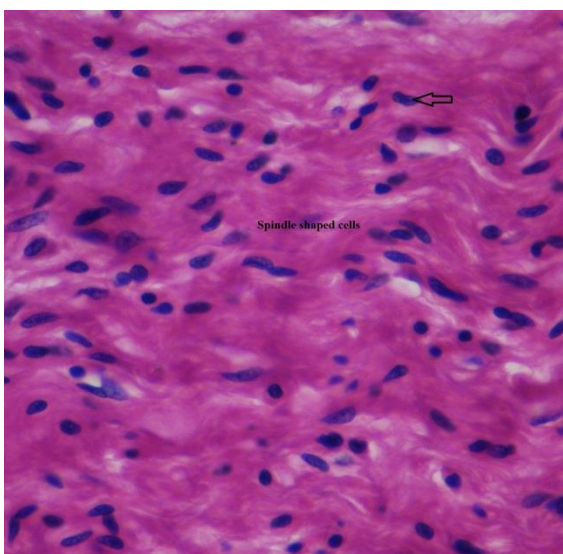


Figure 2: On Histopathological examination section showed elongated, spindle shaped neoplastic cells with poorly defined cytoplasm.

DISCUSSION

Neurofibromatosis or Von Recklinghausen's disease is reported in very early literature, as a disease entity with multiple criteria. It is a benign tumor arising from large and small nerves can occur anywhere in the body except brain and spinal cord. Neurofibromas can present as cutaneous spots, discrete multiple nodules or even as a single large mass and are mainly composed of fibroblasts and Schwann cells.^{2,5} Malignant transformation can take place making the prognosis poor. An inguino-scrotal neurofibroma, as seen in this case is quite rare and only few citations are available in literature.⁴ Most of them are associated with generalized neurofibromatosis, boys affected nearly twice as girls.^{6,7} Through the diagnosis could be made clinically because of the presence of multiple nodules all over the body, it was exactly mimicking an indirect inguinal hernia extending into the scrotum.^{8,9} Other swellings that commonly occur in this region and have to be differentiated are, lipomas of the spermatic cord, which can also mimic an inguinal hernia, angiomyxoma of male genital regions, fibromas, haemangiomas, papillomas and epidermoid cysts.^{7,8} Excision and histo-pathological examination will support the diagnosis, which will show numerous nerve bundles, hyperplastic nerves, proliferating Schwann cells, fibroblasts and perineural cells.^{5,6}

The present case, the mass was a neurofibroma of the anterior abdominal wall in the groin, descending into the thigh. It was separate from the testis, vas deferens and epididymis.¹⁰ Possible origin of this tumor could be from the genital branch of genitofemoral nerve, which innervates the skin on the medial side of the thigh and adjacent scrotal skin. Among available literature our case is similar to the case reported by Milathianakis et al. & Issa MM et al.^{3,11}

CONCLUSION

Neurofibromatosis is an autosomal dominant, benign neoplasm of peripheral nerves. Though it can occur anywhere in the body and present in various forms, it occurs rarely at the genital level especially in the scrotal region.¹² Frozen section microscopic examination should be performed preoperatively if malignancy is suspected. Treatment of choice is surgical, with complete excision of the mass. In our case report, surgery was performed for symptomatic relief and we are reporting this for its rarity.

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