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Case Report

Neurofibroma of Adrenal gland- A rare case report

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ABSTRACT

Neurofibroma of the adrenal glands is rare, most common site of presentation are head and neck region and along the nerves. With the us of computed tomography , magnetic resonance imaging , an ultrasonography for the detection of adrenals masses is increasing. On computed tomography neurogenic tumours frequently appear as a distinct, smooth or lobulated mass. All varieties of neurogenic tumours may exhibit calcification But for confirmation of the lesion histopathological examination and immunohistochemistry is a must. Neurofibroma of adrenals although rare but a possibility.

Key Messages: Immunohistochemistry plays an important role in confirming the diagnosis. IHC helps in definitive diagnosis which helps clinicians with management of the patient.

1. Introduction

Neurofibromas are benign nerve sheath tumors that develop from the perineural and Schwann cells of the normal peripheral nerve; They may appear as solitary lesions or in conjunction with neurofibromatosis type-1 (NF-1), autosomal dominant disease. Most common site being head and neck region, upper extremities or along the nerves. Retroperitoneal, mesenteric and paraspinal location of neurofibromas have also been mentioned.[1] Neurofibroma of the adrenals is an extremely rare disease. Herein, presenting a rare case of 30 years old female diagnosed with neurofibroma of adrenal gland.

2. Case History

A 30 year old female patient presented to Department of Surgery of Vijayanagara institute of medical science (VIMS) Ballari, with complaints of pain and mass per abdomen since five months. CECT of the patient was done and it showed a mass of 7.4 x 6.3cms, which was well defined hypo-enhancing lesion involving lateral limb of right adrenal gland suggestive of benign etiology. Plasma and urine catecholamine levels and serum cortisol levels were within normal limits. Routine hematological investigations were within normal limits. Based on clinical examination and radiological assessment, a diagnosis of right adrenal mass was suggested. Subsequently right adrenalectomy was performed, the specimen was fixed in 10% formalin and was send to Department of Pathology, VIMS Ballari for histopathological examination and immunohistochemistry.

2.1. Grossly

Right adrenalectomy specimen received measuring 8 x 4 x 2 cm, weighing 150gms. External surface was congested and enlarged. The Cut section shows well circumscribed bulging grey yellow areas with interspersed grey white areas.

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Fig. 1: Gross picture of the adrenal

2.2. *Microscopy*

Histological examination of well circumscribed mass shows interlacing fascicle of spindle shaped cells with bland wavy nuclei with sheets of adrenal cells. No necrosis, mitotic activity or infiltrative growth pattern was observed. A diagnosis of Leiomyoma was made. But after immunohistochemistry the tumor cells showed strongly positive for S-100 and negative for SMA and Desmin.

Final diagnosis of Neurofibroma was made.

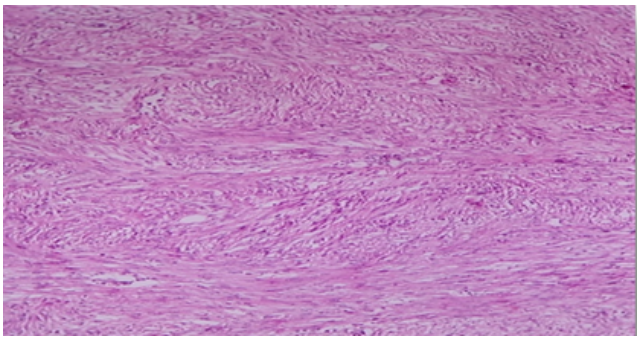

Fig. 2: Low power view, 100x, H&E stain

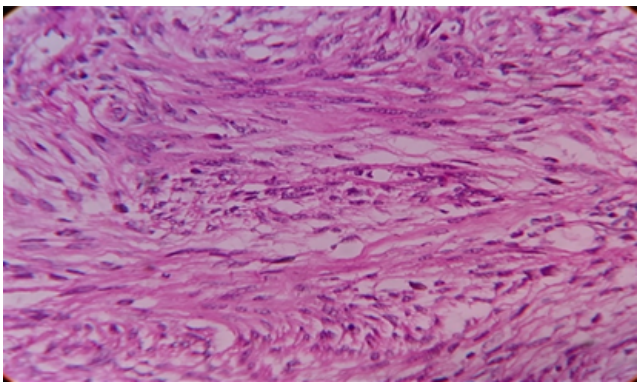

Fig. 3: High power, 400x, H&E stain

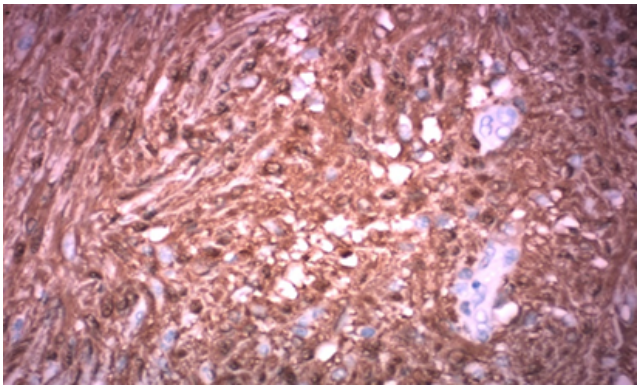

Fig. 4: Immunohistochemistry showing positivity for S-100, 400x

3. Discussion

Any encapsulated nerve in the body can develop neurofibroma, which are classified as follows: 1) solitary neurofibromas, which are well-defined and circumscribed tumors; 2) plexiforms, which are diffuse masses, formed of neural fibers; 3) multiple which are typically linked to hereditary disease; and 4) infiltrating neurofibromas known as diffused neurofibromas.[2]

The nerve sheath is where neurofibromas develop. They demonstrate interspersed growth of nerve sheath cells at histologic assessment. Exhibiting various degrees of myxoid degeneration and thick, wavy collagen bundles

Few cases of solitary neurofibroma, unrelated to neurofibromatosis, have also been reported in the wall of the abdomen, kidney, colon, anal canal, common bile duct and pancrease. The retroperitoneum, paraspinal regions, and adrenals are most frequent sites of neurogenic tumours of the abdomen.[1]

The location of neurofibromas affects the harm they do. Surface distortion is caused by cutaneous tumours, whereas functional compromise and annihilation are possible as a result of deeper masses' compression of nearby organs. The severity of the symptoms is dependent on the size of the lesion, even while the majority are benign, some may cause secondary destruction as a result of the pressure applied.

There is currently no evidence linking solitary neurofibromas to neurofibromatosis. The primary constituents of neurofibromas are Schwann cells, mast cells, fibroblasts, perineural cells, endothelial cells, and collagen. Solitary neurofibromas, which can affect either sexes equally and often develop slowly in the skin of adults between the ages of 20 and 30, are solid, well-circumscribed, differentiated, and non-encapsulated tumours. It is uncommon for neurofibromas to develop in other body regions.[2]

Solitary neurofibroma of the adrenal gland reveals the same morphology as seen at other anatomic sites in the form of spindle-shaped cells with wavy nuclei dispersed

in an extracellular matrix containing variable amounts of mucin and collagen. By immunohistochemistry, these are strongly and diffusely positive for S-100 protein. Many variants of neurofibroma have been reported to date, such as myxoid, hyalinized, epithelioid, Pacinian, pigmented, granular, lipomatous, dendritic cell neurofibroma with pseudo rosettes and neurofibroma with rhabdomyomatous, differentiation.[3]

Adrenal masses are discovered unintentionall in 1-5% of all abdominal CT scans performed; on CT scan, neurofibromas have a homogenous, smooth appearnece with distinct outlines. Because the widespread use of computer tomography(CT), magnetic resonance imaging and an ultrasosography for the evaluation of the diseases with abdominal discomfort. On computed tomography, Neurofibromas appear uniform, smooth, spherical, and have definite contours. They exhibit uniform enhancement with attenuation values of 30–50 HU on contrast-enhanced scans and attenuation values of 20–25 HU on unenhanced scans.[4]

P Gupta et al reported a case of 51 yrs old woman diagnosed with solitary neurofibroma of adrenals.[1] Falahatkar s et al reported a case of 24 years old woman diagnosed with neurofibroma of adrenals.[5] Few literatures also suggest the presentation of neurofibroma at other uncommon site too like Chang CG et al reported a case of abdominal wall neurofibroma presenting as an inguinal hernia.[6] Tsai PJ et al reported a case of neurofibroma of the pancrease body not associated with type 1 neurofibromatosis.[7] Kostakopoulos et al reported a case of solitary neurofibroma of kidney.[8]

Primary treatment modality for many adrenals tumor is considered to be the laproscopic adrenalectomy as it as less post operative complications as compared to open adrenalectomy.

Adrenal glands and the retroperitoneum along the paravertebral sympathetic plexus are where abdominal neurogenic tumours are most frequently found. Radiologically, neurogenic tumours frequently appear as a distinct, smooth or lobulated mass. All varieties of neurogenic tumours may exhibit calcification. On a CT scan, benign and malignant neurogenic tumours look similar, with the exception of the possibility of distant metastatic foci in malignant tumours.

The purpose of imaging is to help create a diagnosis because the majority of adult neurogenic tumours are benign.

Whereas, Histopathological examination and Immunohistochemistry Neurofibroma of adrenals confirms the diagnosis. Neurofibroma not associated with NF-1, which although rare must be considered in differential diagnosis of adrenal gland tumors, as the malignant transformation though rare can occur. Early diagnosis helps the clinician to plan the management and with the patients prognosis.

4. Conflict of Interest

There are no conflicts of interest in this article.

5. Source of Funding

None.

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