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Solitary primary paraganglioma occurring in inguinal canal: An unusual case report

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Abstract

Paragangliomas are commonly seen in adrenal medulla, organs of zuckerkanndl, carotid body. Inguinal region is an unusual site for paragangliomas. We report a case of non-functional extra adrenal inguinal paragangliomas occurring in 22year old female patient, resident of Dakshin Kannada district, who presented with left sided inguinal swelling since 20yrs and lower abdominal pain in the past 3months. The surgeons suspected of inguinal hernia, the radiological imaging revealed mass suspected to be of mesenchymal origin probably arising from nerve sheath. The hematological investigations were within normal limits. On exploration of left inguinal region, a mass was found adjacent to round ligament, the diagnosis of "Extra Adrenal Paraganglioma of the round ligament" was made based on histopathological report.

Keywords: Paraganglioma, inguinal region, extra adrenal, round ligament

1. Introduction

Paraganglia are clusters of neuroendocrine cells associated with the sympathetic and parasympathetic nervous system. Tumors of paraganglia are widely distributed from skull to bottom of the pelvis. The most common localization of paragangliomas is the adrenal medulla and they are usually pheochromocytomas. Extra-adrenal localization is observed in 5-10% of all paragangliomas. The most common extra adrenal paragangliomas occur in carotid bodies, jugular foramen, organs of zuckerkanndl, mesenteric paragangliomas are extremely rare [7].

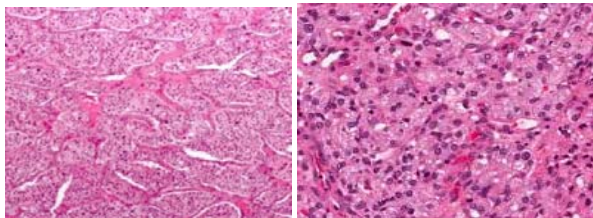
2. Case Report

A 22yr old female presented to our hospital with the complaints of left sided inguinal swelling since 20yrs and lower abdominal pain since 3 months, patient had occasional palpitations. There was no history of vomiting, altered bowel habits and no previous history of similar attacks. The general physical examination revealed mild pallor. Per abdominal examination revealed tenderness and a mass in the left inguinal region of about 4x3cm. Bowel sounds and rectal examination were normal. Examination of other systems were within normal limits except for the respiratory system which showed bilateral basal crepitations on auscultation. The hematological parameters are as follows: Hemoglobin – 9.8gm%, Total WBC count -12,800cells/ cumm, Platelets- 3lakh cells/cumm. The biochemical investigations were within normal limits. Chest X ray was unremarkable. ECG showed sinus tachycardia. An Ultrasonography was advised which showed an inguinal mass suspected to be of mesenchymal origin probably arising from the nerve sheath. On exploration of inguinal region, per operatively a mass was found, which was adjacent to hernia sac, herniorrhaphy and mass was completely resected. The specimen was sent for histopathological examination with a clinical diagnosis of inguinal hernia.

2.1 Histopathological Examination: Gross Examination: single, soft, semi globular well encapsulated grey brown tissue mass measuring 3x2x1cms. The cut section was solid, homogenous grey brown areas seen.



2.2 Microscopy: Sections studied showed tumor composed of large round to polyhedral cells which were separated by fibrovascular septae arranged in cords, trabecular patterns and a characteristic “zellballen” nested appearance with abundant amphophilic cytoplasm. The nuclei were round to oval in appearance with prominent nucleoli.



2.3 Diagnosis: Extra adrenal paraganglioma of the round ligament

Immunohistochemistry was advised but could not be done because of lack of compliance from the patient relatives.

3. Discussion

Paragangliomas are rare neuroendocrine tumors. Neural crest cells give rise to the parenchymal cells of the paraganglia and other elements of the autonomic nervous system. These neural crest cells have the ability to migrate to various regions along the paravertebral and para aortic axis, while remaining in close relation to the sympathetic nervous system. They extend to various places, anywhere from the neck to the base of the pelvis [1].

In rare occasions, paragangliomas have been identified in areas where chromaffin tissue has not yet been characterized, such as the genitor urinary tract, spermatic cord, sacrococcygeal area, anus, renal capsule, broad ligament, ovary, vaginal wall and can only be explained by the migratory property of the neural crest cells [2,9].

Tumors arising from the chromaffin cells of the adrenal medulla are called pheochromocytomas. In our case, tumor was arising from the inguinal canal even though extra adrenal paragangliomas are reported at multiple locations, but no case of paraganglioma in inguinal canal has been reported in literature so far.

Some of the paragangliomas known to originate, store and secrete catecholamines and are hence termed “functional paragangliomas”. While these are easily diagnosed, nonfunctional paragangliomas can pose additional diagnostic dilemmas, thus causing difficulties in the choice of treatment and management options [5, 8].

Hayes *et al.* observed that benign and malignant lesions could not be distinguished based on histopathological findings alone. They opined that distant metastases and local invasion of adjacent organs were the only reliable indicators of malignancy [3]. The tumor in the present case was

interpreted as benign based on the histopathological findings and absence of local invasion.

Malignant extra adrenal paragangliomas are known to metastasize to bone, liver, peritoneum, pelvis, ovaries, cervical lymph node and lung [4]. No evidence of distant metastases was detected in the present case.

4. Conclusion

Paragangliomas can occur in inguinal region; usually, preoperative accurate diagnosis is not possible with imaging modality so diagnosis of paraganglioma pre-operatively is speculative, malignant tumor is a feasible differential diagnosis. Definitediagnosis is possible only after histological evaluation.

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