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URACHAL PARAGANGLIOMA – A RARE CASE REPORT

Pathology

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ABSTRACT

Urachus is a a persistent vestigeal connection between bladder and umbilicus. The lining is usually transitional epithelium which may undergo squamous metaplasia. The development of paragangliomas in urachal tracts is hypothesed to arise from either metaplasia or chromaffin cell migration during embryogenesis. This case report is one of its kind to be reported in literature.

KEYWORDS

Paraganglioma, urachus, Zellballen pattern, synaptophysin

INTRODUCTION

Urachus is a persistent vestigial connection between the dome of urinary bladder and umbilicus. The improper involution of this tubular structure results in the persistence of urachal remnants. The urachus is lined by urothelial epithelium which might undergo metaplasia, leading to the development of neoplasms. The incidence of malignancies arising from urachal tract remnants is 0.5 to 2% [1], majority of which is squamous cell carcinoma. Publications regarding the accurate incidence and presentations of neuroendocrine tumours from urachal remnants are yet to be explored. Our study is among the very few of its type to be reported in Literature.

CLINICAL HISTORY

A 48 years old female patient presented with complaints of abdominal pain for one week. She had no history of haematuria or micturition difficulties. She was not a known case of hypertension, diabetes or cardiovascular diseases. Abdominal MRI of the patient revealed an enhancing solid lesion indenting the urinary bladder dome in midline with obliterated plane of cleavage with underlying bladder wall. It was diagnosed as a Neoplastic lesion – likely of urachal remnant origin with vesical wall invasion(Fig1,2). Abdomino pelvic laparotomy was done and a mass adherent to urinary bladder wall was excised (Fig3).

Gross examination showed urachal tract measuring 7x2.5cm with an adherent solid mass measuring 2x2x1.5cm. External surface was yellowish orange. The cut surface of mass was solid, homogenous, yellowish orange in colour and soft in consistency (Fig4).

Histopathological examination showed a neoplasm arranged in the typical Zellballen pattern surrounded by delicate fibrovascular septa (Fig.5). The individual cells are round to polygonal with abundant granular eosinophilic cytoplasm with vesicular nuclei and coarse chromatin(Fig.6). No lymphovascular invasion was noted. No lining epithelium was noted in the fibrous tract. Immunohistochemistry was done- Chromogranin A showed strong perinuclear dot like golgi positivity in tumour cells (Fig.7). This confirmed the diagnosis of paraganglioma arising from urachal cyst remnants. The postoperative recovery of the patient was uneventful.

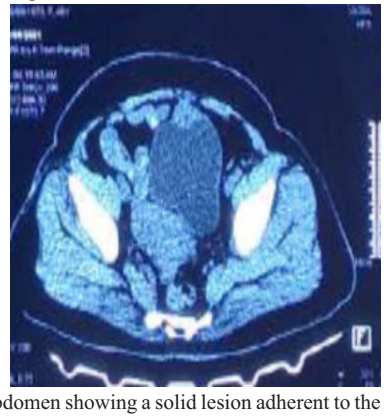

Fig1. CT abdomen showing a solid lesion adherent to the bladder wall with obliteration of cleavage plane

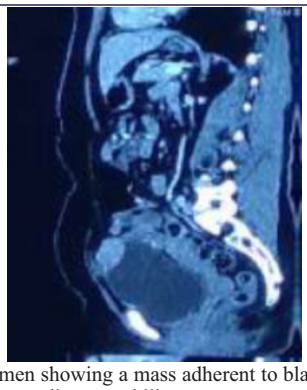

Fig2. MRI abdomen showing a mass adherent to bladder wall with a connecting tract extending to umbilicus

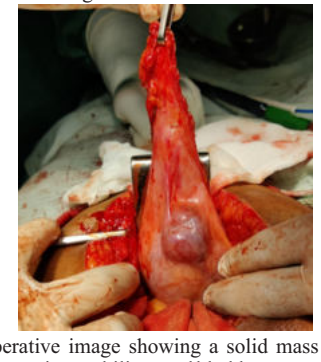

Fig 3. Intra operative image showing a solid mass adherent to the urachal tract connecting umbilicus and bladder.

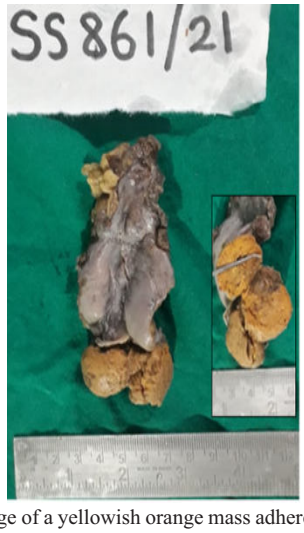

Fig.4 Gross image of a yellowish orange mass adherent to the urachal tract.

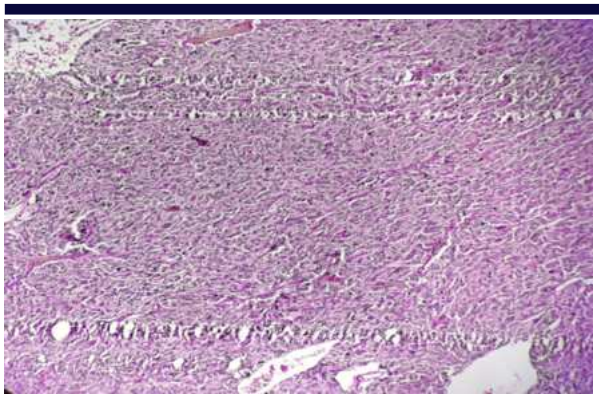

Fig.5 showing typical Zellballen pattern of tumor nests surrounded by fibrovascular septa (10x view, H&E)

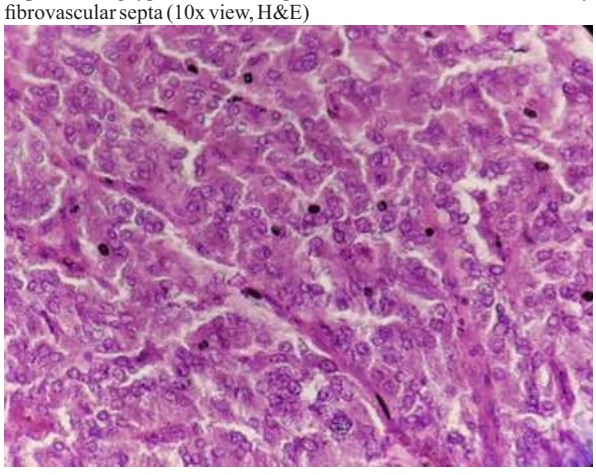

Fig.6 showing tumour nests displaying cells with abundant granular eosinophilic cytoplasm, vesicular nuclei with coarse chromation (40x view, H&E)

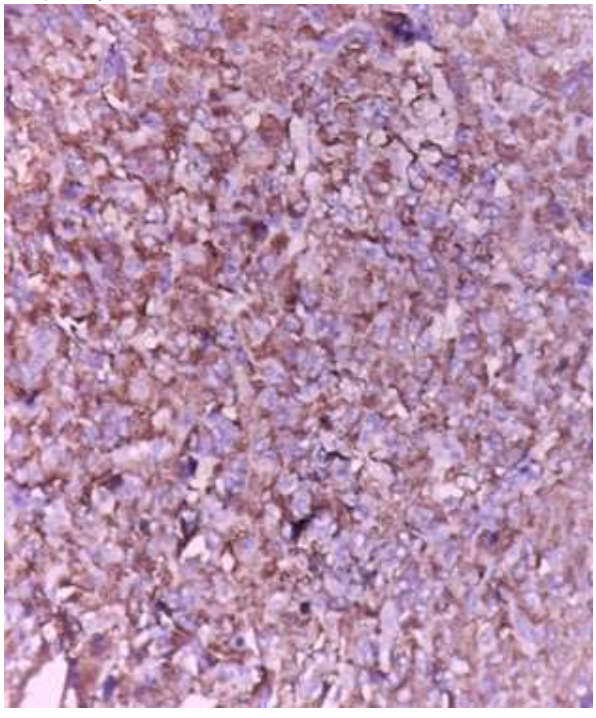

Fig 7- IHC – Chromogranin A showing strong perinuclear dot like golgi pattern of staining (40x view)

DISCUSSION

Urachus is a tubular connection between the urinary bladder and allantois which exists during embryonic development. After birth, it regresses into median umbilical ligament [1]. About 32% of adult autopsies reveal persistence of this urachal tract as urachal remnants. The tract commonly arises from apex of bladder and connects with the umbilicus [2,3]. The canal is lined by transitional epithelium in most cases, surrounded by fibrous stroma. But adenocarcinomas are the commonly encountered malignancies arising from the urachal tract. This is postulated to arise from glandular metaplasia of urachal tract lining [4].Also, there is a possibility of chromaffin cells migrating to the urachal wall during embryogenesis.[5]

Neuroendocrine tumors of bladder are rare neoplasms and neuroendocrine neoplasms of urachus are even rare [3]. Commonly reported symptoms include hematuria, abdominal pain, dysuria and mucosuria (if its associated with mucinous adenocarcinoma). In our study, the patient presented with abdominal pain. No other presenting complaints was recorded. Most of the paragangliomas are non functional. Symptomatic paragangliomas are rare. They express excess of catecholamines resulting in hypertensive episodes, paraoxysmal headaches, micturition syncopes and hematuria. Symptomatic paragangliomas can be easily ruled out by biochemical tests done in urine [6,7].The histomorphology of neuroendocrine tumours resemble their counterparts in any other location. The typical Zellballen pattern surrounded by delicate fibrovascular septa with tumour cells showing characteristic salt and pepper chromatin are typical of neuroendocrine tumors of any location.

Immunohistochemistry highlights the presence of chromogranin granules which show focal perinuclear dot like Golgi pattern of staining. Synaptophysin, GATA 3 and INSM1 are other markers used for confirmation. S100 staining highlights the surrounding sustentacular cells which is crucial in differentiating paragangliomas and carcinoids[8,9]. Adrenergic blockage followed by local excision proves to be curative[10]. But long term follow up is recommended in most cases.[11]

CONCLUSION

Paragangliomas are rare neuroendocrine neoplasms that can develop in various organs. Our case highlights the possible occurrence of paragangliomas even in vestigial organs like urachal cyst remnants. This when diagnosed carries a good prognosis. It should also be considered during differential diagnoses of periurachal tumors.

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