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ADENOCARCINOMA IN A TAIL GUT CYST: A RARE CASE PRESENTATION

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ABSTRACT Tailgut cysts (TGC) are rare congenital benign lesions that occur in children and adults in retrorectal space and represent embryonal remnants of postnatal gut. Adenocarcinoma in a tailgut cyst is rare and less than 20 cases have been reported in English literature. Diagnosis is usually delayed because of non specific symptoms, and is made after biopsy or surgery. We present a case of adenocarcinoma in tailgut cyst which mimicked like sacrococcygeal teratoma in a 34 yr old male patient who presented with complaints of swelling over sacral region since childhood. His FNAC was s/o saccrococcygeal teratoma but after excision of mass via abdominal and perineal route histopathology was s/o adenocarcinoma in a tail gut cyst. Patient received chemotherapy post operatively and was monitored for recurrence with CEA and CA19-9 levels.12 months follow up post chemotherapy is uneventful.

KEYWORDS : Tail gut cyst, adenocarcinoma

INTRODUCTION:-

Tailgut cysts (TGC) are rare congenital benign lesions that occur in children but in adults it is quite rare and occurs in retrorectal or presacral space. Tailgut cysts are more common in females (75-90%) and are mostly asymptomatic. Malignant changes in tailgut cysts are very rare and most common malignancy in patients with tailgut cyst is adenocarcinoma. The first case of a tailgut cyst with malignant epithelial transformation was described by Ballantine in 1931[1].clinical diagnosis is usually delayed due to non specific symptoms and diagnosis is certain only on histopathology. Early surgical excision is the therapy of choice for tail gut cysts due to risk of malignant transformation .We present a case of adenocarcinoma in tail gut cyst in a male from Indian subcontinent.

CASE REPORT-

A 34 years old male presented with complaints of swelling in natal cleft since childhood and pain and discomfort while sitting since few months. On clinical examination, there was single 3X 2 cm size ovoid, firm swelling in sacral region. Per- rectal examination revealed a diffuse swelling posterior to rectum which was mobile. Patient's routine blood investigations were normal. Fine Needle Aspiration cytology (FNAC) from the swelling was suggestive of cystic lesion most likely to be tailgut cyst. USG guided FNAC from sacrococcygeal mass was done showed cytological features s/o sacrococcygeal teratoma and in presence of nuclear pleomorphism and necrosis, malignant transformation cannot be ruled out.

Patient's Magnetic Resonance Imaging (MRI) pelvis revealed large multiloculated solid cystic complex mass lesion in retrorectal and presacrococcygeal space in pelvis and outside pelvis at left side of natal cleft. Intrapelvic component 8x7.6x6.5 cm and extrapelvic component was of size 3 x 3 x 2.5 cms fat planes between mass, rectum, gluteal muscles and adjacent structures were well distinct. No evidence of bone destruction (Fig1 & 2).

Patient was posted for excision of mass. Mass was excised by abdominal and perineal approach. Mass was of 11x9x6 cm with external surface unencapsulated, irregular, firm with areas of haemorrhage and necrosis, variegated in appearance (fig 3 and fig 4). Cut section showed solid and cystic areas with haemorrhage and necrosis. Cystic areas contained yellow gelatinous material. No hair follicles were seen. It also showed solid areas greyish white, firm cartilaginous and fatty areas.

Patient's postoperative period was uneventful and patient was discharged from hospital on day 14 post operative period after suture removal. Histologically it shows multiloculated cyst lined by wide variety of epithelia varied from stratified squamous, transitional, tall columnar type, gastric and mucinous columnar type. Cyst wall showed disorganised fascicles of smooth muscles and numerous blood vessels. It also showed malignant epithelial cells arranged in glandular and tubular pattern along with large areas of hyalinization and calcifications. Individual tumour cells were polygonal with prominent nucleoli showing moderate nuclear pleomorphism and hyperchromasia, mitosis was 2-3/10 HPF (fig 5 and fig 6). Few glands also showed intraluminal necrotic debris. The stroma also showed chronic lymphocytic infiltrate. However, elements derived from other germ layers as skin, neural tissue, heterologous mesenchymal tissue such as cartilage and bone were not identified therefore eliminating possibility of teratoma as was suggested by FNAC report. Patient was followed up and referred to higher centre and was advised Positron emission Tomography (PET) scan and serum CEA and CA 19-9 levels which was normal. Patient was then advised chemotherapy consisting oxaliplatin, capecitabine. Patient completed chemotherapy and 12 months follow up post chemotherapy is uneventful with normal CEA and CA 19-9 levels

DISCUSSION-

Tailgut cysts (TGC) are rare congenital cysts that occur in the retrorectal space and are thought to arise from postanal primitive gut remnants [2]. The retrorectal or presacral space is bounded anteriorly by the rectum, posteriorly by the sacrum, superiorly by the peritoneal reflection, inferiorly the levator ani and coccygeus

muscles, and laterally by the ureters and iliac vessels [3]. TGC have also been referred to as retrorectal cyst hamartoma[4], cyst of postanal intestine, tail gut vestiges, and rectal cyst [5]. TGC should be distinguished from other lesions which may occur in the retrorectal space including teratomas, epidermal cysts, rectal duplication cysts, anal gland cysts, and anal gland carcinomas [5].

Usually during 28 to 35 days human embryo possess a true tail which is caudal to subsequent formation of anus. It normally regresses during 8th week of gestation. Persistence of this postnatal gut in retrorectal space is hypothesized to be origin of tailgut cyst. .

Tailgut cysts are rare congenital anomalies arising from normally regressing postnatal primitive gut. Tailgut cysts are more common in middle aged females. Female to male ratio is 3:1. Malignant transformation in tail gut cyst is very rare and includes adenocarcinoma, carcinoids, neuroendocrine tumours, endometroid carcinoma, adenosquamous carcinoma, squamous cell carcinoma. All patients with tailgut cysts should be assessed for malignancy.

Most of the patients with tailgut cysts are asymptomatic and are usually detected incidentally during investigations. When symptomatic, it presents with mass effect in 50% of patients. It can also present with discomfort on sacral region, low back pain, abdominal pain, swelling over sacral region and obstructive symptoms.

In present case a 34 years old male presented with complaints of swelling over sacrococcygeal region since birth but pain and discomfort over swelling since few months. Patient was investigated and was diagnosed differentially as sacrococcygeal teratoma and tailgut cyst on MRI and FNAC. Patient was operated with excision of the mass lesion by abdominal and perineal approach and histopathology report was suggestive of adenocarcinoma in a tail gut cyst.

Adenocarcinoma in tailgut cyst is mostly misdiagnosed due to unusual symptoms. They can be palpated on per rectal examination as retrorectal mass.

Despite advances in variety of diagnostic techniques like CT scan and MRI precise diagnosis of adenocarcinoma in tailgut cyst can be done by histopathology of biopsy from mass lesion. Imaging modalities such as Computed Tomography (CT) scan provides additional clues in the differential diagnosis [6]. CT scan shows a well-defined homogeneous retrorectal mass with the CT values ranging from water to soft-tissue density [7]. Most TGC can be identified as multiloculated cysts on higher resolution scans. The keratinous or inflammatory debris within a cyst may account for a more solid appearance. Intralesional calcifications or bony destruction of coccyx or sacrum, commonly seen in sacrococcygeal teratomas, are usually absent in TGC. Because calcification is not a feature of TGC, its presence favours the diagnosis of a teratoma or malignancy. MRI imaging reveals a hypointense lesion on T1-weighted images and homogenously hyperintense lesions on T2-weighted images. Although the malignant portions of the tumor are characterized by irregular wall thickening and intermediate signal intensity on both T1- and T2-weighted images, MRI is probably not the best imaging modality to fully differentiate malignant from benign lesions. Differential diagnosis of adenocarcinoma in tailgut cysts includes sacrococcygeal teratoma, rectal duplication cyst, epidermal cyst, metastatic diseases. Adenocarcinoma in tailgut cyst may show increased levels of tumour markers like CEA like other colorectal malignancies.

There no guidelines on definitive treatment of adenocarcinoma found in tailgut cyst as these are very rare tumours. However when found incidentally these lesions should be excised completely. Different types of surgical approaches have been described in the literature for the excision of tailgut cysts. These include a posterior approach, an abdominal approach or a combination of the two. In present case patient was operated for excision of tumour with abdominal and perineal approach. Mass was excised completely and patient was followed up with CEA and CA 19-9 levels and PET scan.

Patients may be subjected to adjuvant radiotheray or chemotheray but the effectiveness and the need for both or either has not been validated. Chemotherapy and radiotherapy on the basis of serum CEA levels has been advocated.

Saurabh Chabra et al reported a case of adenocarcinoma in the tail gut cyst who after surgical excision did not receive any adjuvant therapy and did not report recurrence on 12 months follow up.[8]

Graadt van roggen et al reported a case of adenocarcinoma in tail gut cyst with CEA positive cells on histopathology, and administered 50.4 cGy of local radiotherapy post operatively.CEA levels remained negative on follow up for 4 yrs but subsequently patient became symptomatic and had increase in CEA levels, suggesting local recurrence. Patient declined further intervention and died soon afterwards.[9]

Our patient was subjected to chemotherapy of oxaliplatin and capecitabine based regimen for 6 months following referral to higher centre and follow up of 12 months post chemotherapy is uneventful.

CONCLUSION—

Tailgut cysts are rare congenital anomalies arising from normally regressing postnatal gut. Adenocarcinoma in tailgut cysts are rare and usually found incidentally and require high suspicion for diagnosis.

Malignant changes in tail gut cyst are very uncommon and may present in long standing cyst as in our case. But definitive diagnosis can only be done on histopathology.

There are no definitive guidelines for treatment of adenocarcinoma in tailgut cysts as these are rare tumours. Surgical excision of these tumours and subsequent adjuvant chemoradiation based on CEA levels and adequacy of resection remain the cornerstone of management. Overall though Adenocarcinoma in tailgut cyst have poor prognosis.

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LEGENDS OF FIGURES:-

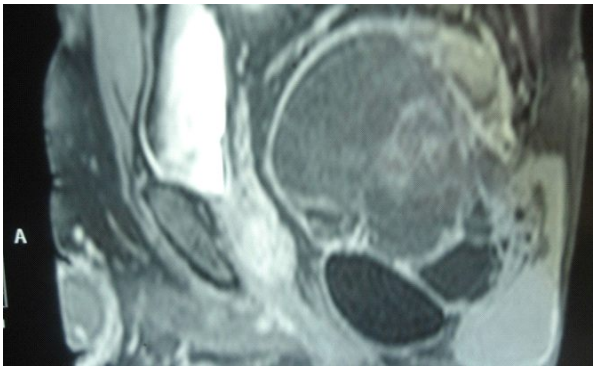

FIGURE 1

Large multiloculated solid cystic complex mass lesion in retrorectal and presacrococcygeal space in pelvis and outside pelvis at left side of natal cleft. Intrapelvic component 8x7.6x6.5 cm and extrapelvic component was of size 3 x 3 x 2.5 cms.fat planes between mass, rectum, gluteal muscles and adjacent structures were well distinct. No evidence of bone destruction.

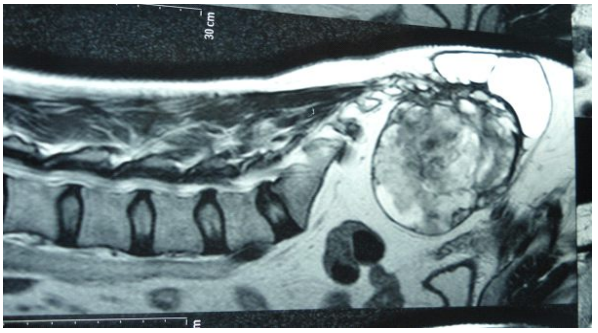

FIGURE 2

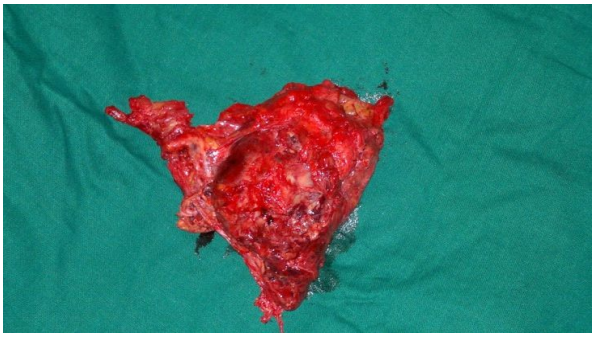

FIGURE 3

Mass was of 11x9x6 cm with external surface unencapsulated, irregular, firm with areas of haemorrhage and necrosis, variegated in appearance. Cut section showed solid and cystic areas with haemorrhage and necrosis. Cystic areas contained yellow gelatinous material. No hair follicles were seen. It also showed solid areas greyish white, firm cartilaginous and fatty areas.

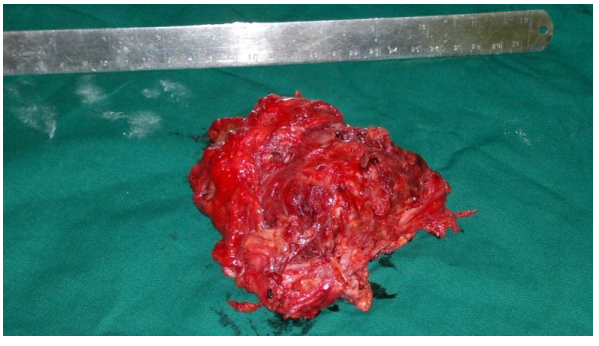

FIGURE 4

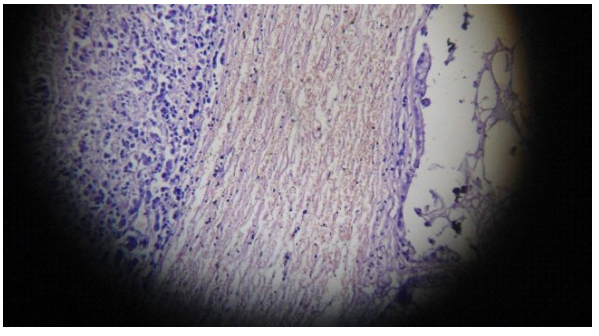

FIGURE 5

Histologically it shows multiloculated cyst lined by wide variety of epithelia varied from stratified squamous, transitional, tall columnar type, gastric and mucinous columnar type. Cyst wall showed disorganised fascicles of smooth muscles and numerous blood vessels. It also showed malignant epithelial cells arranged in glandular and tubular pattern along with large areas of hyalinization and calcifications. Individual tumour cells were polygonal with prominent nucleoli showing moderate nuclear pleomorphism and hyperchromasia, mitosis was 2-3/10 HPF. Few glands also showed intraluminal necrotic debris. The stroma also showed chronic lymphocytic infiltrate

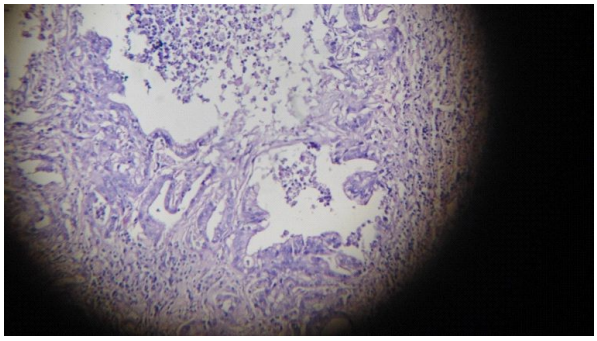

FIGURE 6

REFERENCES-

1. Ballantine EN. Sacrococcygeal tumours. Adenocarcinoma of a cystic congenital embryonal remnant. Arch Pathol 1931; 14:1–9.
2. Cho BC, Kim NK, Lim BJ, et al. A carcinoembryonic antigen-secreting adenocarcinoma arising in tailgut cyst: clinical implications of carcinoembryonic antigen. Yonsei Med J 2005; 46:555-61.
3. Jarboui S, Jarraya H, Mihoub MB, et al. Retrorectal cystic hamartoma associated with malignant disease. Can J Surg 2008; 51:E115-6.
4. Tampi C, Lotwala V, Lakdawala M, et al. Retrorectal cyst hamartoma (tailgut cyst) with malignant transformation. Gynecol Oncol 2007; 105:266-8.
5. Au E, Anderson O, Morgan B, et al. Tailgut cysts: report of two cases. Int J Colorectal Dis 2009; 24:345-50.
6. Schwarz RE, Lyda M, Lew M, et al. A carcinoembryonic antigen-secreting adenocarcinoma arising within a retrorectal tailgut cyst: clinicopathological considerations. Am J Gastroenterol 2000; 95:1344-7.
7. Krivokapic Z, Dimitrijevic I, Barisic G, et al. Adenosquamous carcinoma arising within a retrorectal tailgut cyst: report of a case. World J Gastroenterol 2005; 11:6225-7.
8. Saurabh chabra, Wise S, Maloney-patel, Rezac C, Poplin E."JOURNAL OF GASTROINTESTINAL ONCOLOGY".2013 Mar 4(1)97-100
9. Graadt van Roggen,K Welvaart,A de Roos,G J A Offerhaus, P C W Hogendoorn, "Adenocarcinoma arising within a tailgut cyst: clinicopathological description and follow up of an unusual case." Journal of clinical pathology 1999; 52:310-312