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FETUS IN FETU – A RARE CASE REPORT OF AN ABDOMINAL LUMP

General Surgery

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ABSTRACT

Mature cystic teratomas, or dermoid cysts, are part of the family of non-seminomatous germ cell tumors composed of somatic cells from at least two germ layers. They usually present as a pelvic mass arising from the gonads. The presence of teratomas in the retroperitoneum is a rare phenomenon occurring due to sequestration of germ cells from the yolk sac during migration. We present a case of an abdominal lump in a 20 year-old female which was provisionally diagnosed as a teratoma on imaging. A formal exploration was done and the lump, weighing approximately 7kg, was excised. Lump was confirmed as a mature teratoma of subtype Fetus in Fetu on histopathology. This rare congenital anomaly has been defined as the presence of a malformed and parasitic fetus in the body of its twin. Its incidence is about 1:500,000 live births. Retroperitoneal teratoma is rare and even rarer is the possibility of a Fetus in Fetu. This case is being reported with relevant salient features and is discussed in the light of available literature due to its distinctive characteristics.

KEYWORDS

Abdominal Lump, Mature Teratoma, Dermoid Cysts, Fetus In Fetu, Retroperitoneal Tumor.

INTRODUCTION

Primary retroperitoneal neoplasms comprise only 0.1–0.3% of all retroperitoneal tumors, out of which 1–11% are mature dermoid cysts. [1] Extra-gonadal teratomas are quite rare and are derived from aberrant migration of embryonic cells. They are found in the midline structures such as anterior mediastinum, sacrococcygeal region, retroperitoneum, pineal gland, neck and abdomen. [2] Cases are reported usually in infancy or young females. The most common site in the retroperitoneum is left suprarenal region in adults. [3] Retroperitoneal teratomas are twice as common in females as in males. [2,3]

Dermoid cysts are of 4 types- sequestration, implantation, tubular and teratomatous. Teratomatous dermoids are cystic swellings that contain an array of developmentally mature and solid tissues. They are usually lined by squamous epithelium showing dermal papillae with sebaceous gland, hair and sweat follicle and contains sebaceous cheesy material, bone and teeth. Most are benign, but any tissue type can undergo malignant transformation. The presence of immature tissue increases the likelihood of malignant transformation.

Retroperitoneal teratomas are asymptomatic in one-third of patients. They can grow to a considerable size before becoming symptomatic. The symptoms are due to compression of adjacent structures resulting in vomiting, constipation, lumbar back pain, abdominal distention and urinary complaints. Systemic symptoms may also be present such as fever, chills, night sweats, and weight loss.

Fetus in fetu (FIF) is a terminology used for a teratomatous mass, which shows similarity to a developing fetus. The true embryogenesis of FIF is not adequately understood. One theory describes FIF to be the result of uneven blastula division leading to a small proportion of cells being separated forming a diamniotic, monochorionic and monozygotic teratomatous twin.[4] The other theory suggests FIF a mature teratoma with tissue from different germ layers arising from the vestiges of the wolffian / müllerian ducts or the pronephric / mesonephric tubules as it correlates with their midline and paramedian location. [5]

CASE REPORT

A 20 year old female, hailing from an obscure tribal area, noticed a progressively slow-growing lump in abdomen since the last 12 years. There were no associated complaints. No relevant surgical, medical or family history was noted.

On examination, a large mass was palpable covering the epigastric, both hypochondrium and umbilical regions with a downward shift of the umbilicus. The lump was large, non-tender, non-mobile, firm in consistency and not moving with respiration. Upper margin was imperceptible but the lower margin was well defined. (Figure 1- A, B)

The patient was anemic on admission. A plain abdominal radiograph showed calcification in the epigastrium. Ultrasonography of abdomen suggested a complex mass with anechogenic and echogenic components along with internal vascularity and multiple septations. The mass measured 18 x 16 x 16 cm and was displacing all adjacent organs. Contrast Enhanced Computed Tomography of the abdomen suggested a dermoid cyst (teratomatous dermoid). (Figure 1- C, D, E) The solid cystic lesion was causing mass effect by displacing the pancreas anteriorly and resultant thinning of the parenchyma. The common bile duct was compressed with mild central and peripheral intra-hepatic biliary dilatation with compression of the portal vein, superior mesenteric artery and vein. The bowel loops were displaced inferiorly and kidneys postero-laterally. Serum alpha fetoprotein was normal.

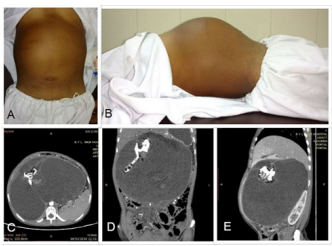

FIGURE 1: A and B: Patient Preoperative picture depicting the

abdominal lump in epigastric, hypochondrium and umbilical region with lower margin visible and umbilicus displaced inferiorly.; **C, D and E:** Preoperative CT scan depicting lump with calcification and adjacent mass effect displacing the pancreas anteriorly and resultant thinning of the parenchyma. The common bile duct is compressed with mild central and peripheral intra-hepatic biliary dilatation with compression of the portal vein, superior mesenteric artery and vein. The bowel loops are displaced inferiorly and kidneys postero-laterally.

Intra-operatively, a large retroperitoneal mass spanning the entire abdominal cavity and measuring approximately 26 x 22 x 25 cm was noticed. There were dilated and engorged vessels surrounding the mass. Due to close proximity to major vessels, decision was taken to aspirate the cyst and reduce its size before any curative en-masse excision. An anterior cystotomy was done and fluid within was aspirated, taking care to prevent spillage and contamination of the peritoneal cavity. About 1800 cc of fluid was aspirated. Inspite of aspiration the size continued to be large and hence a decision taken to remove the contents piecemeal. About 7 kg of sebaceous cheesy material mixed with hair, teeth, bony components, skin appendages and a placenta-like structure was taken out. The remnant cyst wall was excised but posterior cyst wall was left behind due to dense adhesions with major vessels. The inner lining of the remnant cyst wall was cauterized and a drain was kept in the cavity. Patient's post-operative recovery was uneventful. Regular povidone iodine washes were given through the drain until the returning fluid was free from any debris. Drain was removed on post op day 6 and patient discharged on day 11.

Final histopathology report confirmed a mature cystic teratoma with no evidence of malignancy or immature elements. (Figure 2 A, B, C, D). On follow up imaging a near complete excision was noted (Figure 2 E, F).

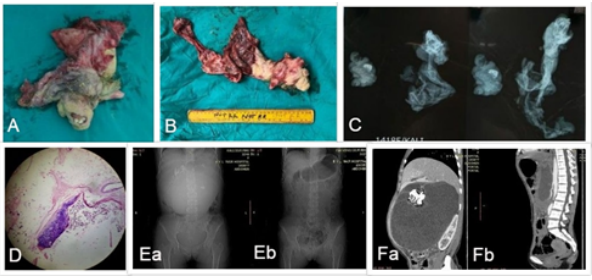

Fig 2: A and B: Specimen depicting tooth, placenta like structure and appendages; **C:** Specimen X ray depicting axial vertebral column and tooth; **D:** Histopathology view of the specimen showing cartilage, bone and epithelial keratin squames. **Ea:** Pre op plain coronal view with visible epigastric calcification; **Eb:** Post op view without calcification; **Fa:** Pre op contrast enhanced sagittal view with lump compressing the superior mesenteric artery; **Fb:** Post op view with marked decrease in lump and prominent flow through the superior mesenteric artery.

DISCUSSION

Willis states "A teratoma is a true tumor or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises." [6] FIF may be a very highly differentiated form of dermoid cyst, itself a highly differentiated form of mature teratoma. [7] An early example of the phenomenon was described in 1808 by George Young 8 and the term was coined by Johann Meckel. [4] FIF frequently inhabits the retroperitoneal region. The presence of an axial skeleton helps differentiate the lump from similar fetiform teratomas.

Symptoms of retroperitoneal teratoma are variable, but in benign cases there is rarely an alteration in the general condition. In malignant forms the initial clinical picture may be normal, but there are often symptoms or disturbances due to compression. Risk of malignancy ranges from 6.8% to 36.3% and increases with age, male sex, and presence of immature tissues. [9] Malignant teratomas appear to be the only extrahepatic tumors which can produce serum alpha fetoprotein, this substance disappear 8 to 10 days after tumor removal and reappear in case of a recurrence. [5] However, as benignity cannot be ascertained, the tumor must be removed surgically. Tissue adherence, which has been observed with malignant and benign lesions, may hinder complete removal or require extended surgery.

Lastly, even though FIF is a fascinating terminology and gives the impression that there is a baby incorporated in another baby, its management and that of a mature teratoma is essentially the same - complete excision. The diagnosis of FIF has important clinical implications as most FIF after resection can ensure a permanent cure.

CONSENT OF PATIENT

The patient signed the informed consent for surgery and the publication of this case before the intervention.

CONFLICT OF INTEREST

The authors declare that they have no competing interests.

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