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Sacrococcygeal Yolk Sac Tumor – An Unusual Case Report

A.Srujana [1] , P.Premalatha [2], I.V.Renuka [3] , Rayappa Reddy Thumma [4]
[1234]*Department of Pathology, NRI Medical college, Dr. NTR University of Health Sciences, India*

Abstract: *A 2 year old female child presented with a tender visible lump progressing into an abscess in the sacral region. MRI revealed a huge sacral mass pushing the rectum anteriorly. Serum alpha fetoprotein was abnormally elevated. Histhopathological examination of the surgical specimen suggested a sacrococcygeal yolk sac tumor.*

Keywords: *Alphafetoprotein,MRI, sacral mass, sacrococcygeal yolk sac tumor*

I. Introduction

Extragonadal germ cell tumors commonly occur in mediastinum, suprasellar regions, sacrococcyx and retroperitoneum in the descending order of frequency. Majority of sacrococcygeal tumors are benign teratomas [1] These tumors however have the potential for malignant degeneration. Malignancy is usually limited to a single element, a yolk sac tumor [2]. This tumor may less commonly be present in "pure" form. Malignant germ cell tumors account for 3% of childhood neoplasms [3]. Although, most germ cell tumors in children originate in the gonads, the most common primary site for YST is the sacrococcygeal region [4]. Sacrococcygeal yolk sac tumor is extremely rare with only a few case reports found in literature.

II. Case Report

A 2 year old female child presented with a tender visible lump of 6 cm x 5 cm progressing into an abscess in the right buttock of two months duration for which incision and drainage was done. The child had a normal birth history with no significant past medical history. Abdominal and pelvic magnetic resonance imaging (MRI) demonstrated a large 6.5 x 5cm solid mild enhancing mass located in precoccygeal region, displacing rectum anteriorly with extension outside pelvis in posterior aspect of coccyx (Fig-1). There was a significant mass-effect on adjacent organs. Serum alpha fetoprotein level was 1012 ng/ml (normal range: 0-8 ng/ml). Ultrasonography (USG) of the ovaries was normal. Retroperitoneal lymph nodes were not enlarged. Her X-ray chest was clear.

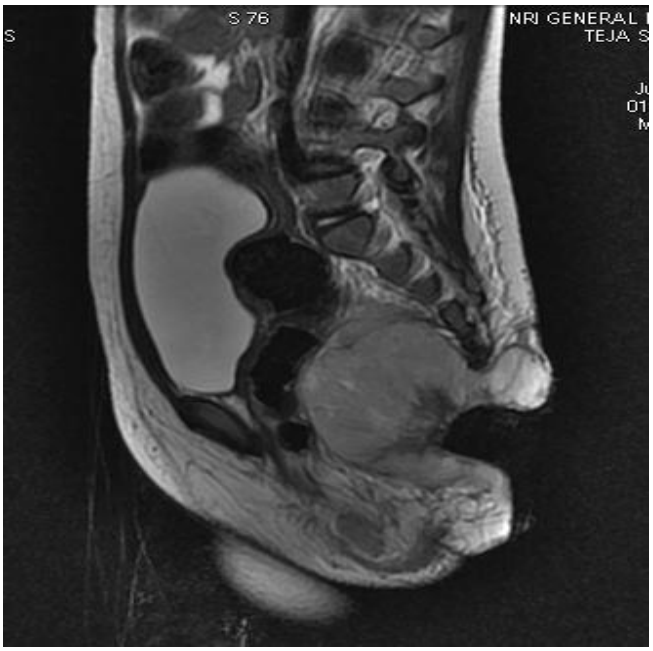

Fig 1- Abdominal and pelvic magnetic resonance imaging (MRI) showing a large 6.5 x 5cm solid mild enhancing mass located in precoccygeal region, displacing rectum anteriorly.

2.1 Gross: Received multiple grey white friable necrotic tissue bits and membranous bits altogether measuring 12x8cm. Cut section – solid, grey white with foci of myxoid areas.

2.2 Microscopy: Histopathological examination showed a primitive teratoid malignant tumor exhibiting predominant reticular growth pattern with microcysts and loose textured myxoid areas surrounded by cords and sheets of tumor cells, some in vague papillary and glandular patterns as well as perivascular disposition of tumor cells resembling Schiller – Duval bodies). Neoplastic cells were relatively large, with pale eosinophilic cytoplasm and some showing hyaline droplets with round to oval vesicular nuclei. There were large areas of tumor necrosis with areas of benign chondroid and osteoid differentiation (heterologous elements)(Fig-2; A-F). The patient completed 5 cycles of neoadjuvant chemotherapy which included bleomycin, etoposide and cisplatin following which the AFP levels returned to normal.

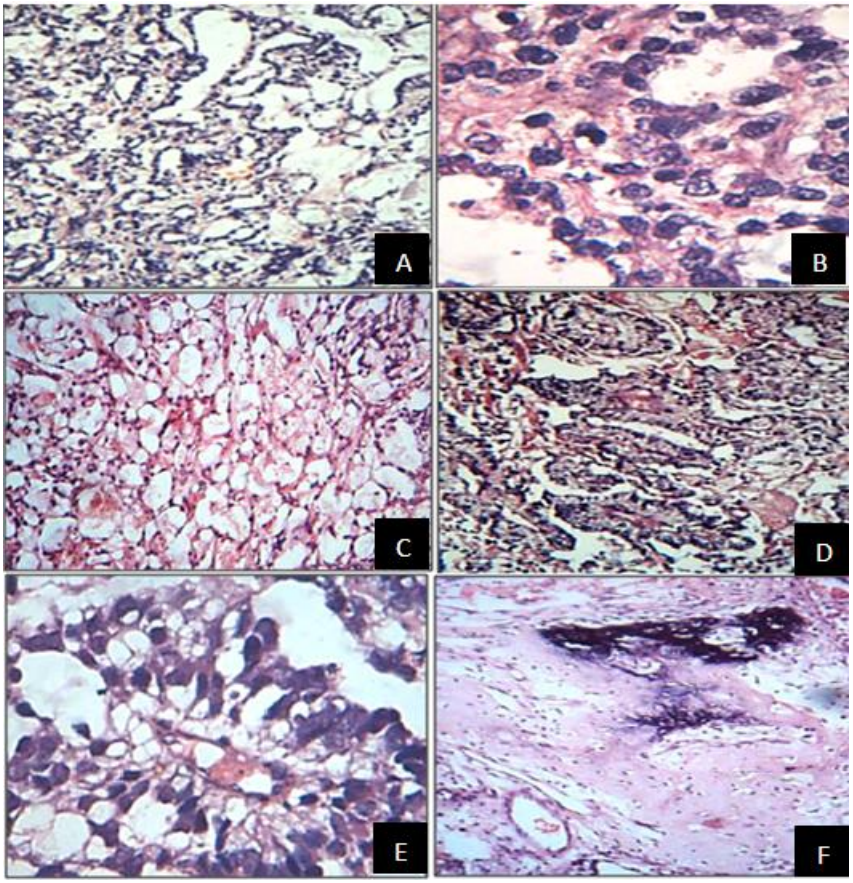

Fig 2: Histopathological patterns of YST. (A)Reticular growth pattern; H & E x100 (B) Glandular; H & E x100 (C)Polyvescicular vitelline pattern; H & E x100 (D) Endodermal sinus pattern; H & E x100 (E) Schiller- Duval body ;H & E x400 (F)Heterologous elements; H & E x100

III. Discussion

Germ cell tumors are a heterogeneous group of neoplasms. Their origin in extragonadal location is due to abnormal migration of germ cells in the embryo. Sacrococcygeal germ cell tumors in infants and neonates are nearly always primary. 75%-90% occur in females. Teratoma is the most common malignant germ cell tumor in the sacrococcygeal region. Yolk sac tumors may occur as primary (de novo) in infants or emerge in a residual teratoma after incomplete resection after two years of life or still rarely as a metastatic tumor from a ovarian primary. Sacrococcygeal YST develops exclusively in children less than 2 years of age [4,5,6].

Imaging of sacral tumors in children plays a significant role in identifying the position, contents and invasion. YST is often complicated with hemorrhage, necrosis and cystic degeneration. Thus on CT and MRI the signal density is heterogeneous. Honeycomb-like change is an imaging characteristic of YST [7]. Obscure boundary between tumor and surrounding tissue, sacral invasion and metastases are signs of malignancy. AFP determination is useful in the diagnosis, to monitor the results of therapy and detects metastases and recurrence after therapy.

Gross examination of YSTs is predominantly solid, soft, grey white. Cystic degeneration, necrosis and hemorrhage are often present.

As described and illustrated by Teilum[8] in his writings on this tumor microscopic patterns of YST are numerous. Several different patterns are usually admixed. They are characterized by the intermingling of epithelial and mesenchymal elements in a specific organoid fashion. Microcystic, glandular-alveolar and

papillary formations are common. Many of the cystic spaces are lined by flattened, endothelium-like layer of cells. The stroma can be cellular, spindle shape and reminiscent of smooth muscle. Perivascular Schiller-Duval bodies, which are almost always present in sizable samples but may be absent in limited material, as in biopsies, are the most distinctive features of yolk sac tumor. Periodic acid Schiff positive intracytoplasmic hyaline and extracytoplasmic droplets are consistently seen in yolk sac tumors.

Immunohistochemically, these tumors stain for AFP. However, immunohistochemical assay may be negative for AFP in some cases. Most of the immunoreactivity is seen diffusely or in granular fashion throughout the cytoplasm of the tumor cells [9].

The treatment of malignant sacrococcygeal GCTs, such as primary yolk sac tumor is dependent on the extent of disease. Local disease is best managed surgically, while advanced tumor stages benefit best from adjuvant platinum based chemotherapy which warranties a good prognosis. Survival rates using these strategies are higher than 80% [10].

IV. Conclusion

Extragonadal yolk sac tumors are extremely rare. This is a case of pure form of sacrococcygeal yolk sac tumor with no prior history of any surgeries in the past thus excluding the possibility of residual teratoma progressing into a malignancy. Inspection of both ovaries was done to exclude a gonadal primary. Hence it was reported as a primary sacrococcygeal yolk sac tumor (YST). Neoadjuvant chemotherapy was given followed by wide local excision surgery.The prognosis of primary sacrococcygeal YST is good with possible complete remission when compared to metastatic sacrococcygeal YST.

However as these tumors are highly malignant with early metastasis and rapid invasion to adjacent organs, adjuvant chemotherapy plays a very important role even after complete resection of tumor.

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