CASE REPORT

Mature Cystic Teratoma with Neuroendocrine Tumor— Case Report

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Abstract

Presacral tumors are extremely rare tumors in adults.Such lesions are often found incidentally.The strange presentation of lesions in the pre-sacral space and the ambiguity of its clinical presentation makes it difficult to establish its diagnosis as the first possibility. Given that several germ lines converge in this presacral space, it is possible to find a variety of lesions, one of the rarest being neuroendocrine tumors. We present a case of 34 year old female diagnosed with pre-sacral mass that was surgically resected, with histologic studies showing a mature cystic teratoma with neuroendocrine tumor.

Key Words

Presacral Tumor, Neuroendocrine Tumor, Sacrococcygeal Region

Introduction

Presacral tumors are a rare and heterogeneous group of tumors occurring in the potential space between the rectum and the sacrum. They are estimated to account for about 1 in 40,000 hospital admissions.These tumors are usually more common in the female sex (29-71 years age), with up to 66% of them being of benign origin. [1]

In the United States from 1994 to 2009 the incidence of neuroendocrine tumors increased from 2.48 to 5.85 cases per 100,000 per year, in Europe and Asia the incidence is significantly lower ranging from 1.1 to 3.24 cases per 100,000 per year. Despite the above, in the presacral region they are extremely rare.[2]

Presacral tumors range from simple benign cysts to complex malignant masses.Primary carcinoid tumor (well-differentiated neuroendocrine tumor) of the bone involving the sacrum is extremely rare

Case Report

We present a case of 34 year old female, previously healthy with no past medical or surgical history, presenting with the complaints of insidious,non radiating pain in lower abdomen (on & off) for 6 months which was aggravated since last 15 days. She had no history of nausea, vomiting,fever, hematochezia/melena, abdominal distension, weight loss, or trauma. Her bowel and bladder habits were normal, and she had no family history of malignancy. On examination patient was afebrile, hemodynamically stable, and fully conscious. Her abdomen was soft and non-tender with normal bowel sounds, and digital rectal examination revealed extensive bulge in the dorsolateral rectal wall.

CT scan of Abdomen & Pelvis revealed large soft tissue attenuation mass (8.3 X 8.8 cms) in sacral region with erosion of underlying sacrum with mass effect in the form of displacement of rectum on left lateral aspect with variable degree of enhancement of contrast in the lesion with necrotic core within.

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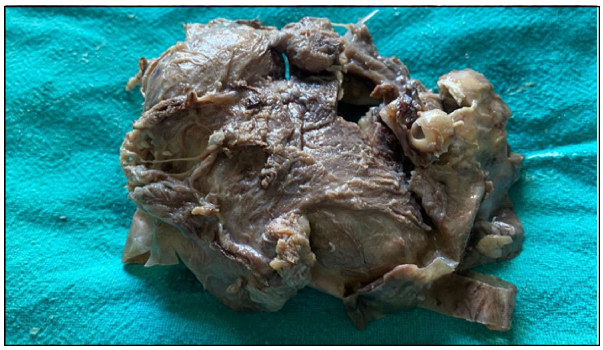

Fig 1: Gross appearance of the specimen showing the external shiny surface of the mass.

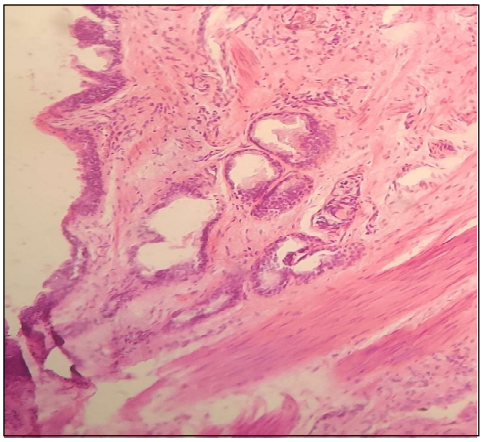

Fig 2: Tumor lined by stratified squamous epithelium withintra-cystic accumulation of flakes of keratin. Presence of mucin secreting glands is also seen.

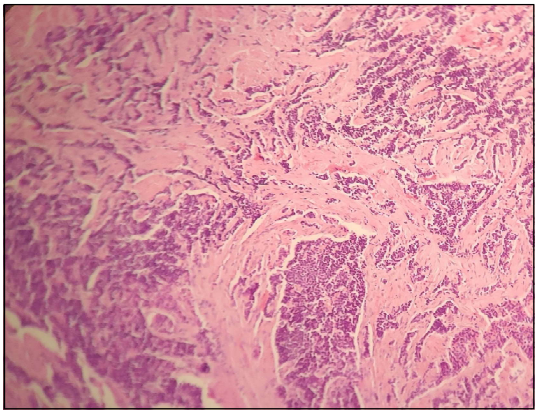

Fig 3: Lobules and cords of mildly pleomorphic cells with salt and pepper chromatin with scanty mitotic activity. Focally these tumor cells are seen infiltrating in a single line pattern in the muscle.

The patient successfully underwent a surgery following which the specimen was received in our Department.

Grosslythe external surface was smoothand on cut section, solid and cystic areas were identified (Figure 1)

Microscopically multiple sections taken from pre-sacral mass showed tumor lined by stratified squamous epithelium with intra-cystic accumulation of flakes of keratin.

Cyst also showed adnexal structures, mucous secreting glands, cartilage. There were thick smooth muscle fascicles in the cyst wall admixed with foci of mature adipose tissues.

Respiratory epithelium, collection of macrophages and foci of calcification was also identified.

Immature cartilage and neural elements were not identified. All these features were consistent with Mature Cystic Teratoma.

In addition, there were lobules and cords of mildly pleomorphic cells with salt and pepper chromatin with scanty mitotic activity. Focally these tumor cells were seen infiltrating in a single line pattern in the muscle. Mitosis was scanty. (Figure 2 & 3)

On Immunohistochemistry, Sall4, p63, S100 and Desmin stained Mature Teratoma. NSE and Synaptophysin highlights neuroendocrine component with Ki 67 = 1. GFAP was negative.

AnIHC proven diagnosis of Mature Cystic Teratoma with well differentiated Neuroendocrine tumor was made.

Discussion

Pre-sacral tumors were first reported by Emmerich in the mid-19th century. Given its rarity, its exact percentage of presentation is currently unknown; however, some articles record an incidence of approximately 1 case in 40,000.[1]

In a review of the literature, only 15 reported cases of neuroendocrine tumors of primary origin in the presacral space were found, of which up to 60% of the cases were female and the majority were associated with sacro-octogenic teratomas or cysts of the posterior intestine.[3]

Teratomas usually occur in the gonads but 1–5% occur inextra gonadal sites.[4]

Teratomas are congenital. It is thought that, during embryological migration, some of the cells arrest and survive in an extragonadal location, mainly at mid-line sitesthat is why 1–5% of cases are found in extra gonadal sites. In order of frequency these sites are: anterior mediastinum, retroperitoneal, sacrococcygeal and intracranial.[5,6]

Extra gonadal teratomas are generally benign with an approximate 25% risk of malignant transformation.[7] Neuroendocrine carcinoma within a teratoma is extremely rare.

One of the theories that arises about the origin of neuroendocrine tumors in the presacral region is described by Kim *et al*.,[3] who suggests that these tumors derive from embryological remnants of the posterior intestine, since histopathological similarity has been found between these cells and tumors.[3] One of the most in frequent presenting lesions, in this location, are neuroendocrine tumors.

The association of primary carcinoid tumor with sacrococcygeal teratoma also is a rare condition.The symptoms of sacrococcygeal teratoma usually are non-specific.Chronic low back pain isthe most common presenting symptom.[8]

The prognosis of pre-sacral tumors generally depends on the histological nature of the lesion and on complete surgical resection.Although the prognosis is good after complete surgical resection in a benign tumor, malignant tumors remain a problem and the prognosis is poor.Complete resection of the tumor is crucial and can prevent recurrence. However, after a successful operative intervention there is no standard regimen for the use of chemotherapy or radiation.

Recurrence rate usually low (7%) and a survival rate reaching 100%.[9]

Conclusion

Primary NETs in thepresacral region are very rare and the clinical presentation is often ambiguous making its diagnosis difficult. An IHC is necessary to confirm the diagnosis.In terms of treatment, surgical excision is needed to remove the primary tumor.

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