

## Presacral immature teratoma with carcinoid in a young adult male: A rare case presentation

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### ABSTRACT

Presacral region is defined as potential space delineated by the rectum anteriorly, sacrum posteriorly, peritoneal reflection superiorly, and perineal muscles inferiorly. Tumors within the presacral space are uncommon with an incidence of 1:40,000. Presacral teratomas in adults are very rare. We, herein, present a case of 39-year-old male who was admitted with chief complaint of per rectal bleed. A large presacral mass was present which on histopathological examination was found to be immature teratoma with neuroendocrine differentiation.

**Key words:** Presacral, Immature, Teratoma, Tumors

Presacral region is defined as potential space delineated by the rectum anteriorly, sacrum posteriorly, peritoneal reflection superiorly, and perineal muscles inferiorly [1]. Tumors within the presacral space are uncommon with an incidence of 1:40,000 [2]. The most common tumor in presacral space is chordomas [3]. The tumors in presacral space may arise within tail gut cysts and teratomas suggesting the congenital nature of these tumors [3,4]. Teratomas within the presacral space most commonly occur in infancy and have an incidence of 1 in 30,000–43,000 live births with a 3–4:1 female-to-male ratio [2,5,6]. Rarely, do presacral teratomas present in adulthood. We, herein, present a case of 39-year-old male who was admitted with chief complaint of per rectal bleed. A large presacral mass was present which on histopathological examination was found to be immature teratoma with neuroendocrine differentiation.

### CASE REPORT

A 39-year-old male was hospitalized with the chief complaint of constipation and per rectal bleeding for past 4–5 months. These symptoms were on and off initially but now progressed. The patient had not taken any treatment of the same in the past. On examination, the patient was bit pale with the normal vital signs. On per rectal examination, a large firm, nontender mass was found between the rectum and sacrum. A complete hemogram was performed which showed a hemoglobin of 5.1 g/dl with all other parameters being normal. Rest of the routine investigations was normal. Magnetic resonance imaging was done which showed the presence of a large well defined solid cystic mass in the presacral space with solid component showing

intense heterogeneous enhancement (Fig. 1). The mass was seen compressing and displacing the rectum anteriorly (Fig. 2). The possibility of malignant teratoma was being suggested.

Complete excision of the mass was done and sent for histopathological examination. Grossly, the mass was gray white to gray brown measuring 8 cm × 7 cm × 3.5 cm. Cut section of this mass showed partial solid areas with multiple cysts ranging in size from 0.5 to 2 cm in diameter. These cysts were filled with pultaceous material (Fig. 3). Histopathological examination revealed the presence of immature teratoma with neuroendocrine differentiation possibly carcinoid (Figs. 4–6). An immunohistochemistry panel was applied which showed immunoreactivity for neuron-specific enolase. However, glial fibrillary acidic protein was found to be negative (Fig. 7). Diagnosis of immature teratoma with carcinoid was made and the patient was advised for complete surgical excision.

### DISCUSSION

Presacral space contains multiple embryological remnants and is a site for the development of various types of tumors. Various tumors or tumor-like lesions including teratoma, chordoma, myxopapillary ependymal, paraganglioma, schwannoma, liposarcoma, tail gut cyst, and metastatic tumor can occur in this area with increased incidence. Chordomas are the most common among them [3]. Presacral teratomas in adults are very rare. The first reported case of a presacral teratoma in an adult was published by Emmerich in 1847 and was included as one of six cases reviewed by Kiderlen, in 1899 [7]. Wishnia *et al.* in their study, in 2008, demonstrated fewer than 120 reported cases of isolated adult presacral teratoma [8]. Of those patients whose sex was reported (92 of 104 patients), 70% were female and 30% were male.

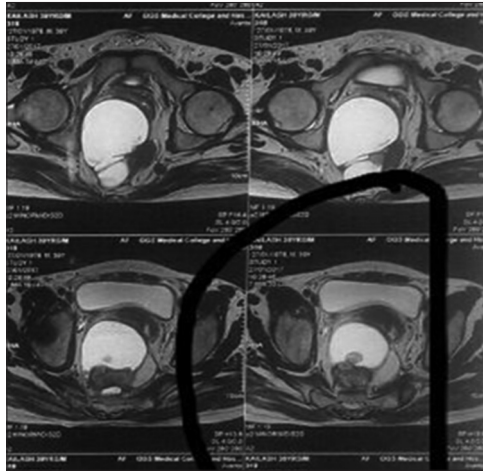


Figure 1: Large well defined solid cystic mass in the presacral space with solid component showing intense heterogenous enhancement

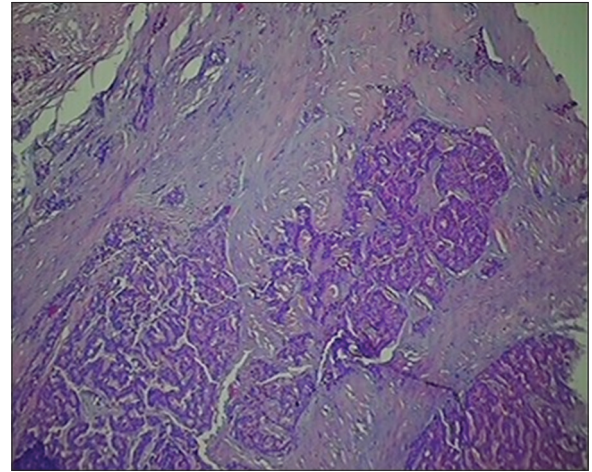


Figure 4: Low power view of the tumor arranged in trabecular pattern with the presence of myxoid stroma

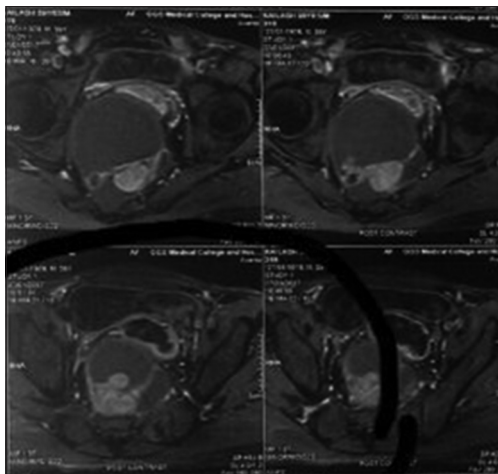


Figure 2: This mass was seen compressing and displacing the rectum anteriorly

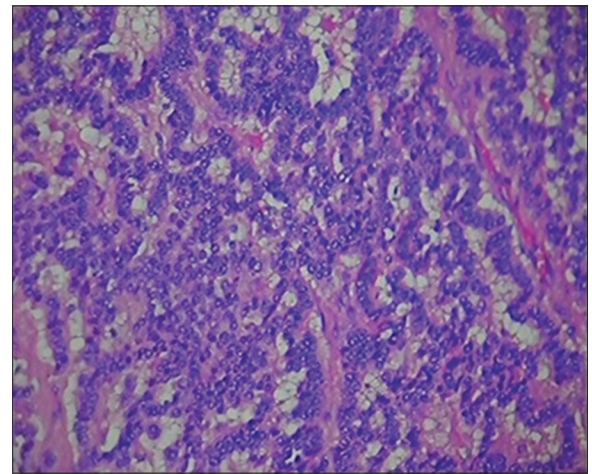


Figure 5: High power view showing individual tumor cells having round to oval nuclei having granular chromatin and mild amount of cytoplasm



Figure 3: Grossly, the mass was gray white to gray brown measuring 8 cm x 7 cm x 3.5 cm. Cut section showed partial solid areas with multiple cysts ranging in size from 0.5 to 2 cm in diameter

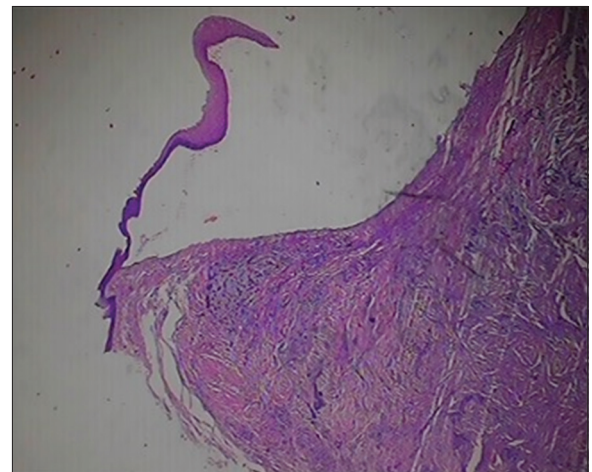
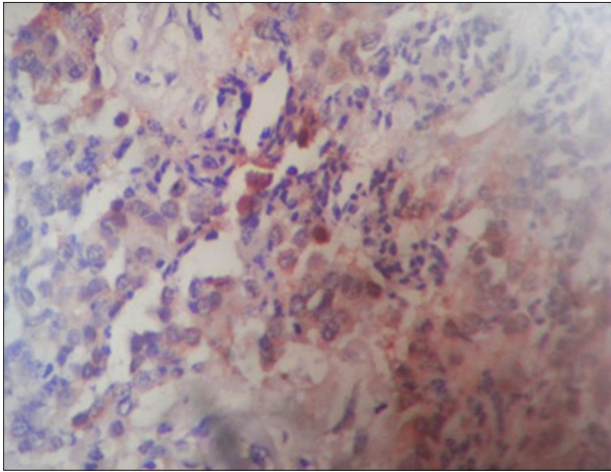


Figure 6: Squamous component of the teratoma is seen as lining of stratified squamous epithelium

Macroscopically, teratomas are partially cystic masses filled with gelatinous fluid or keratin. Microscopically, they contain a variety of cell types derived from more than one germ layer [9]. Teratomas can be classified into three histologic categories: Mature, immature, and malignant. Mature teratomas have benign epithelial-lined cysts, mature cartilage, and/or muscle. Immature teratomas contain primitive mesoderm, endoderm, or ectoderm admixed with the mature elements. Malignant teratomas contain

frank malignant tissue which may or may not be of germ cell origin [6]. Carcinoid tumors within the presacral space are rare





**Figure 7: Immunohistochemistry showing neuron-specific enolase positivity**

and most often represent the direct extension or metastatic spread from adjacent rectal carcinoids [2,10]. Dujardin *et al.* found 20 cases of primary neuroendocrine carcinoma in the presacral space reported in the literature in 2009 [11]. Spada *et al.*, in 2010, stated that only 28 cases of carcinoid tumors of the presacral region are described in the medical literature and of these 14 were associated with tail gut cysts. The rest was either associated with teratomas or with no other peculiar anomalies [12].

These tumors are managed by surgical resection. The prognosis of mature teratoma is good while malignant teratoma is poor with a higher tendency to recur and metastasize [5,6]. The 5 years survival rate for presacral neuroendocrine carcinoma with metastasis has been reported to be 20–30% [11].

## CONCLUSIONS

Carcinoid tumors in presacral region are rare. Malignant components within a teratoma need to be differentiated from

primary and metastatic tumors. These tumors should be included in the differential diagnosis of presacral mass and close follow-up is required for early diagnosis and management of recurrence or metastasis.

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