Case Report

Sacro-coccygeal teratoma in adult: Two rare case reports and review of literature

Narendra Kumar, Divya Khosla, Ritesh Kumar, Uma Nahar Saikia[1], SK Singh[2]

Department of Radiotherapy and Oncology, Regional Cancer Centre, Departments of [1]Histopathology, and [2]Urology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Abstract

The sacrococcygeal area is the most common site of extragonadal teratomas in infants, but is a challenge to make clinical as well as radiological diagnosis in adults. We herein describe two cases of sacrococcygeal teratoma (SCT) in adult. The clinical, radiological and histopathological characteristics of both the cases with their outcome are described with review of the literature. The standard care for SCTs is complete surgical resection of the tumor. The presence of malignant transformation is associated with a less favorable outcome.

Key words: Adult, mature, sacrococcygeal, teratoma, treatment

Submission: 09-05-2013 **Accepted:** 11-01-2014

Introduction

Sacrococcygeal teratoma (SCT) is the most common congenital neonatal tumor but rarely seen in adults. Prevalence of SCT varied from 1/14,900 to 1/40,000 live births in various series and occurs more often in girls; with female to male ratio of 3-4:1.[1-4] Most of the SCTs are cystic and benign and only 1-2% are malignant.[5] The cysts may be filled with serous fluid, mucoid, or sebaceous material and lined by true epithelium. Radiological imaging is helpful in the diagnosis of these lesions and in delineating their extent. Treatment consists of complete surgical resection and coccygectomy. Prognosis of SCT depends on the extent of surgical resection and histopathological findings of tumor (benign or malignant). The two cases reported here illustrate the variable clinical course of this rare tumor.

Address for correspondence: Dr. Ritesh Kumar,
Department of Radiotherapy and Oncology, Regional Cancer Centre,
Postgraduate Institute of Medical Education and Research,
Sector 12, Chandigarh - 160 012, India.
E-mail: riteshkr9@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijabmr.org
	DOI: 10.4103/2229-516X.136803

Case Reports

Case 1

The first case is about a 37-year-old male patient who presented with a history of gradually increasing low back pain for 3 years, occasional episodes of difficulty in micturition with no significant change in bowel habits. Routine clinical examination of the chest and abdomen was normal. Digital rectal examination (DRE) revealed an extra-luminal cystic mass in the pre-sacral region with normal rectal mucosa. Neurological examination was unremarkable. Chest roentgenography, complete blood cell count, biochemical profile were within normal limits. Alpha-fetoprotein (AFP), carcinoembryonic antigen, lactate dehydrogenase (LDH) and β-human chorionic gonadotropin (β-HCG) levels were also normal. Contrast enhanced magnetic resonance imaging (MRI) revealed a large mass lesion measuring 13.5 cm × 7 cm × 5 cm with epicenter in sacrum consisting of large pre-sacral and retro-sacral soft-tissue component with fluid levels in lesion [Figure 1]. The lesion had intracanalicular component compressing cauda equina nerve roots. The patient underwent gross total excision of tumor with coccygectomy. On gross examination, the surgical specimen looked like a biloculated cyst containing sebum with partly hemorrhagic and necrotic appearance. Smaller cysts contained clear serous fluid. Microscopic examination showed tumor containing all three germ layers characteristic of a teratoma. The cyst was partly lined by stratified squamous epithelium containing hair follicles and sebaceous

glands and other areas revealed pseudostratified ciliated columnar epithelium (endodermal derivation). The stroma was composed of neuroglial tissue (ectodermal derivation); adipose tissue and bone (mesodermal derivation) [Figure 2a and b]. A small fragment showing pancreatic tissue was also identified along with abundant fibrocollagenous tissue. The solid region was composed of irregular nests of neoplastic glands with morphological features of a moderately differentiated adenocarcinoma, infiltrating the surrounding stroma with desmoplastic reaction, hence confirming the diagnosis of adenocarcinomatous transformation in a mature teratoma. There was no lymphovascular invasion.

In view of aggressive histopathological features to take care of microscopic disease patient was planned and given post-operative radiotherapy, a dose of 45 Gy in 25 fractions at 1.8 Gy per fraction over 5 weeks followed by adjuvant combination chemotherapy with bleomycin, etoposide and cisplatin for three cycles. No significant radiotherapy and chemotherapy induced toxicity encountered. The patient was then kept on regular follow-up. At 6 months after completion of treatment, routine follow-up imaging with MRI showed multiloculated cystic lesion with solid components in the pre-sacral region extending to perirectal space and bilateral ischiorectal fossa suggestive of recurrence of disease. Patient was referred for surgical opinion, but patient refused any further active treatment.

Case 2

The second case is a 21-year-old female patient who presented with the complaints of a mass in the sacrococcygeal area for 2 years and gradually increasing in size. No complaints of lower limb weakness or bowel and bladder dysfunction. On examination, a huge swelling approximately 20 cm × 20 cm in size was present in the sacrococcygeal area. It was of variable consistency and not fixed with skin. DRE revealed an extra-luminal cystic mass in the pre-sacral region. Tumor markers (AFP, LDH, β-HCG) were within normal limits. MRI scan showed a large (25 cm × 25 cm) well-defined cystic lesion in sacrococcygeal area [Figure 3]. Intra-operatively, there was a large cystic mass in sacrococcygeal region with extension to the lower third of the rectum and adherent to the coccyx. The rectum was dissected off the pre-sacral fascia and the tumor was dissected along with coccyx. Patient underwent total excision with pelvic floor repair and diversion sigmoid colostomy. Histopathological examination revealed a tumor with intricate admixture of ectodermal (epidermis, sebaceous glands, squamous and hair), mesenchymal (cartilage, adipose tissue, blood vessels, nerves and skeletal muscle) and endodermal components (respiratory epithelium, intestinal epithelium) respectively [Figure 2c and d]. A small focus of neuroglial tissue was also seen and no immature components were seen. Overall features were those of mature teratoma.

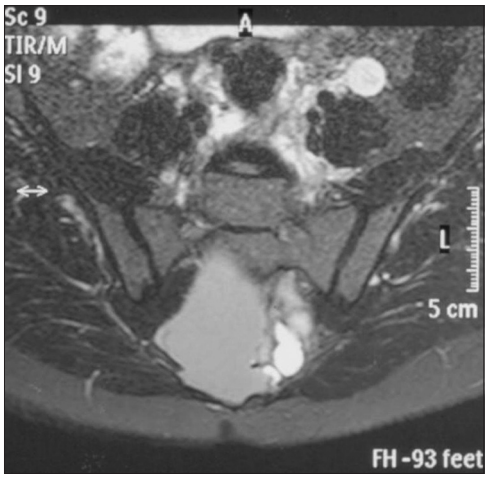

Figure 1: Magnetic resonance imaging showing a large mass lesion in pre-sacral area with large soft-tissue component

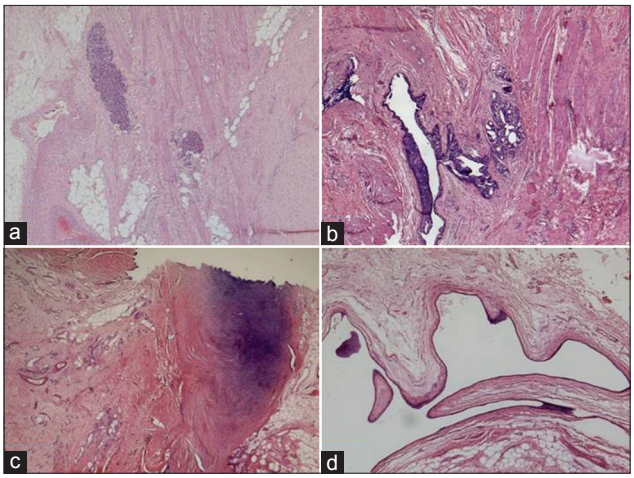

Figure 2: a) Photomicrophotograph showing lobules of fat, smooth muscle and acini (H and E, ×2.5). (b) Glands lined by columnar and squamous lining surrounded by smooth muscle (H and E, ×4). (c) Mature cartilage with surrounding glandular structures. (d) Large cyst lined partly by flattened to columnar to squamous lining (H and E, ×2.5)

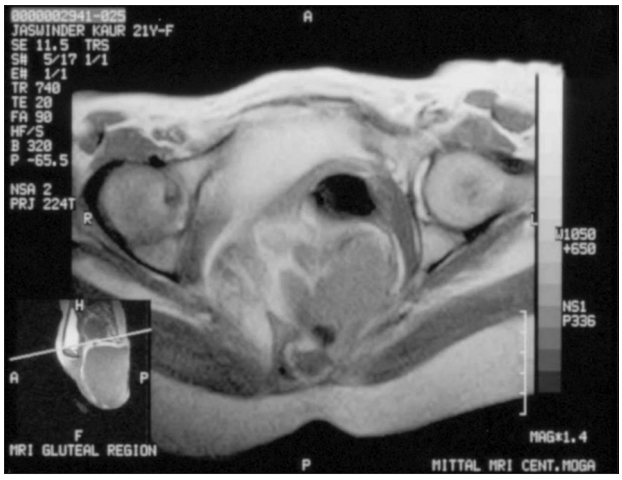

Figure 3: Magnetic resonance imaging showing a large well-defined cystic lesion in sacrococcygeal area

The patient recovered without showing any post-operative complication. In view of mature teratoma without any aggressive features and complete surgical excision patient was kept on regular follow-up. At 20 months of follow-up patient is both clinically and radiologically disease free.

Discussion

Teratomas are germ cell tumors commonly composed of multiple cell types derived from one or more of the three germ cell layers. SCTs are rare in adults with only a few published cases in the literature.[6-10] Many theories have been postulated about the development of SCT. These include derivation from totipotential cells of the primitive knot or Hensen's node, embryonic entities that contribute to the gonadal ridge and eventually end up in the coccygeal region. Other theories include non-sexual reproduction of germ cells within the gonads or in extragonadal sites; "wandering" germ cells of non-parthenogenetic origin left behind during the migration of embryonic germ cells from the yolk sac to gonad; or origin in other totipotential embryonic cells.[7] A theory of incomplete twinning has also been proposed.[11]

Most of the adult SCT present as intrapelvic masses, whereas on the contrary in neonates, 90% of SCT are externally visible. Symptoms may be related to mass effects or bulk of the tumor, such as low back pain, bowel or urinary symptoms, venous engorgement of the lower limbs and lower extremity motor power losses. Calcifications in the coccygeal region in the roentgenogram or an anterior displacement of the rectum in the barium enema are findings suggestive of SCT.[8] computed tomography and MRI are the most significant tools to characterize the mass, to evaluate the intrapelvic extension and relationship to other structures. Macroscopic examination commonly shows partially cystic lesions.[8] Both of our patients had cystic tumor of varying consistency and size. Microscopic features include the presence of derivatives of more than one germ layer. SCTs are classified into three categories based on their histopathological features: entirely mature adult-type tissue, immature and malignant.[9] Mature teratoma (also referred to as benign teratoma) contains obvious epithelial-lined structures as well as mature cartilage and striated or smooth muscle. Immature teratomas contain primitive mesoderm, endoderm or ectoderm mixed with more mature elements. Malignant teratomas have frankly malignant tissue of germ cell origin in addition to mature and/or embryonic tissues.

The differential diagnosis of SCT in adults includes chordoma, meningocele, giant cell tumor of sacrum, pilonidal cysts, rectal duplication cysts or anal gland cysts osteomyelitis of sacrum, fistula with pre-sacral extension and abscess formation, post-injection granuloma and tuberculosis.[10] The standard treatment for all primary SCTs is surgical excision. Because these tumors are contiguous with the coccyx, which may contain the nidus of totipotential cells, it is recommended that the coccyx be removed. Failure to remove the coccyx has been associated with a high risk of recurrence.[1] If the tumor is histologically benign (mature tissues only) or immature teratoma without frankly malignant tissue, complete excision is adequate. For malignant teratomas, surgical excision alone is inadequate and patients should receive additional treatment with chemotherapy and/or radiotherapy. In view of the rarity of these tumors, there has been no standard recommendation for the use of chemotherapy or radiation.

Conclusion

SCT should be considered in the differential diagnosis of a pelvic mass in adults. Long-term survival is possible with complete removal of the tumor and benign histology. The presence of malignant transformation and incomplete excision are associated with a less favorable prognosis and outcome.

References

1. Schropp KP, Lobe TE, Rao B, Mutabagani K, Kay GA, Gilchrist BF, *et al*. Sacrococcygeal teratoma: The experience of four decades. J Pediatr Surg 1992;27:1075-8.
2. Pauniaho SL, Heikinheimo O, Vettenranta K, Salonen J, Stefanovic V, Ritvanen A, *et al*. High prevalence of sacrococcygeal teratoma in Finland-A nationwide population-based study. Acta Paediatr 2013;102:e251-6.
3. Golas MM, Gunawan B, Raab BW, Füzesi L, Lange B. Malignant transformation of an untreated congenital sacrococcygeal teratoma: A amplification at 8q and 12p detected by comparative genomic hybridization. Cancer Genet Cytogenet 2010;197:95-8.
4. Swamy R, Embleton N, Hale J. Sacrococcygeal teratoma over two decades: Birth prevalence, prenatal diagnosis and clinical outcomes. Prenat Diagn 2008;28:1048-51.
5. Al-Essa AA, Malik TA, Baghdadi MK, El Tayeb AA. Adult sacrococcygeal teratomas. Saudi Med J 2004;25:367-9.
6. Ghosh J, Eglinton T, Frizelle FA, Watson AJ. Presacral tumours in adults. Surgeon 2007;5:31-8.
7. Audet IM, Goldhahn RT Jr, Dent TL. Adult sacrococcygeal teratomas. Am Surg 2000;66:61-5.
8. Ng EW, Porcu P, Loehrer PJ Sr. Sacrococcygeal teratoma in adults: Case reports and a review of the literature. Cancer 1999;86:1198-202.
9. Luk SY, Tsang YP, Chan TS, Lee TF, Leung KC. Sacrococcygeal teratoma in adults: Case report and literature review. Hong Kong Med J 2011;17:417-20.
10. Miles RM, Stewart GS Jr. Sacrococcygeal teratomas in adult. Ann Surg 1974;179:676-83.
11. Isaacs H. Germ cell tumors. In: Isaacs H, editor. Tumors of the Fetus and Newborn. Philadelphia: WB Saunders; 1997. p. 15-38.

How to cite this article: Kumar N, Khosla D, Kumar R, Saikia UN, Singh SK. Sacro-coccygeal teratoma in adult: Two rare case reports and review of literature. Int J App Basic Med Res 2014;4:122-4.

Source of Support: Nill. **Conflict of Interest:** None declared.