Giant Mature Sacrococcygeal Teratoma in an Adult - a Case Report

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Citation

A Sheikh, S Solanki, F Solanki, S Hussain, D Sharma, D Sharma, V Raina. *Giant Mature Sacrococcygeal Teratoma in an Adult - a Case Report*. The Internet Journal of Surgery. 2008 Volume 21 Number 1.

Abstract

Teratoma is a congenital germ cell tumor, composed of a wide diversity of tissue, containing all three germ cell layers. The sacrococcygeal area is the commonest site of extragonadal teratomas in infants, but presents as a rare diagnostic dilemma in adults. We present a rare case of giant mature sacrococcygeal teratoma in an adult female patient.

CASE REPORT

A 17-year-old female belonging to rural area presented with complaints of a mass in the sacrococcygeal area since birth and gradually increasing in size. There were no neurological complaints of lower limb weakness or bowel and bladder dysfunction. On examination, a huge swelling approximately 25 x 25cm in size was present in the sacrococcygeal area (Figure I).

Figure 1

Figure I Clinical photograph of sacrococcygeal teratoma



It was of variable consistency and not fixed with skin. On per rectal examination, the rectal mucosa was free from masses. The patient was anemic (hemoglobin: 8g%) with normal sugar and urea level. His chest X-ray was normal. Alpha fetoprotein was 1 IU/ml (N: 0.55-2.64), LDH was 307 IU/l (N: 114-240 IU). CT scan showed a huge (25 x 25cm) fatty mass with some soft tissue and few calcifications with disfigurement of the sacrum (Figures II and III).

Figure 2

Figure II CT scan image of sacrococcygeal teratoma



Figure 3

Figure III CT scan image of sacrococcygeal teratoma



The mass was completely excised from the sacral approach (Figure IV).

Figure 4

Figure IV Excised sacrococcygeal teratoma (macroscopic view)



Histo-pathological examination showed predominant adipose tissue with presence of muscle, fibrocollagenous tissue, epidermis, dermis and mature cartilage, i.e. mature teratoma.

DISCUSSION

When the current focus on sacrococcygeal teratoma is on prenatal assessment, fetal intervention [1] and outcome after surgery in the pediatric age group [2], sporadic cases of sacrococcygeal teratoma in the adult age group come as a surprising rarity. A survey of literature reveals few case reports and even fewer series of cases. [3-8] In fact, a recent comprehensive review could find only 92 published cases in the literature, confirming its rare incidence. [9]

Its incidence has been reported to be 1:87,000 with a female to male ratio of 10:1. [10] Its rarity is further compounded by varied symptoms, which are due to its size, compression of pelvic viscera or complications like infections, bowel obstruction, pelvic abscess, pilo-nidal sinus, degeneration/fistulas, or malignancy. A 20-30% risk of secondary infection and a 1-12.5% risk of malignant degeneration has been calculated. [11] Most adult teratomas described are small, found in female patients and rarely present externally. [12]

The differential diagnosis includes anterior sacral meningocele, anal duct or gland cyst, necrotic rectal leiomyosarcoma, extraperitoneal adenomucinosis, cystic lymphangioma, pyogenic abscess, neurogenic cyst, and necrotic sacral chordoma. [13] Preoperative biopsy should be avoided as it can cause tumor spread, or abscess formation and should not be performed if the tumor is potentially resectable. [3] Good appropriate imaging in the form of CT or MR scan remains the cornerstone of decision making in its management.

The treatment of sacrococcygeal teratomas is mainly surgical. Increasing experience with retro-rectal/presacral tumours and better imaging has resulted in evolving classifications and algorithms for their management. [14-17] The approach depends on the peculiarities of each case (size, location and invasion of the nearby viscera), with most teratomas amenable to resection from posterior sacral approach. Resection of the coccyx is considered necessary to prevent local recurrence [12]; however, it is logical in case of its involvement by the neoplastic mass or suspected malignant transformation. Anterior or combined approaches are rarely required for tumours invading the viscera. Prognosis is directly related primarily to local control, which may be difficult to achieve for malignant lesions.

As late occurrence of malignancy following resection of a histologically mature sacrococcygeal teratoma has been reported [18], our patient has been kept on 3-monthly follow-up.

References

 Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, Adzick NS. Sacrococcygeal teratoma: prenatal assessment, fetal intervention, and outcome. J Pediatr Surg 2004; 39(3): 430-8.
Derikx JP, De Backer A, van de Schoot L, et al. Longterm functional sequelae of sacrococcygeal teratoma: a national study in The Netherlands. J Pediatr Surg 2007; 42(6): 1122-6.

3. Jao SW, Beart RW Jr, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors. Mayo Clinic experience,

1960-1979. Dis Colon Rectum 1985; 28(9): 644-52.

4. Ng EW, Porcu P, Loehrer PJ Sr. Sacrococcygeal teratoma in adults: case reports and a review of the literature. Cancer 1999; 86(7): 1198-202.

5. Hobson KG, Ghaemmaghami V, Roe JP, Goodnight JE, Khatri VP. Tumors of the retrorectal space. Dis Colon Rectum. 2005; 48(10): 1964-74.

6. Gan JC, Zhang S, Suo RZ, Zhang ZX. Diagnosis and treatment of sacrococcygeal teratoma in adults: analysis of 17 cases. Zhonghua Yi Xue Za Zhi 2008; 88(31): 2191-4. [Article in Chinese]

7. Mentes BB, Kurukahvecioğlu O, Ege B, et al. Retrorectal tumors: a case series. Turk J Gastroenterol 2008; 19(1): 40-4.

8. Coco C, Manno A, Mattana C, et al. Congenital tumors of the retrorectal space in the adult: report of two cases and review of the literature. Tumori 2008; 94(4): 602-7.

9. Kostas B, Zisis G, Christos T, Isaak TK, Vrakas G, Vrakas X. Sacrococcygeal teratoma in adults: Report of a case and literature review. The Internet Journal of Surgery

2007, Volume 11, Number 2. 10. Al-Essa AA, Malik TA, Baghdadi MK, El Tayeb AA.

Adult sacrococcygeal teratomas. Saudi Med J 2004; 25(3): 367-9.

11. Chêne G, Voitellier M. Benign pre-sacral teratoma and

vestigial retrorectal cysts in the adult. J Chir (Paris) 2006; 143(5): 310-4. [Article in French]

12. Miles RM, Stewart GS Jr. Sacrococcygeal teratomas in adult. Ann Surg 1974; 179(5): 676-83.

13. Dahan H, Arrivé L, Wendum D, Docou le Pointe H, Djouhri H, Tubiana JM. Retrorectal developmental cysts in adults: clinical and radiologic-histopathologic review, differential diagnosis, and treatment. Radiographics. 2001; 21(3): 575-84.

14. Lev-Chelouche D, Gutman M, Goldman G, Even-Sapir E, Meller I, Issakov J, Klausner JM, Rabau M. Presacral tumors: a practical classification and treatment of a unique and heterogeneous group of diseases. Surgery. 2003; 133(5): 473-8.

15. Buchs N, Taylor S, Roche B. The posterior approach for low retrorectal tumors in adults. Int J Colorectal Dis 2007; 22(4): 381-5.

16. Woodfield JC, Chalmers AG, Phillips N, Sagar PM. Algorithms for the surgical management of retrorectal tumours. Br J Surg 2008; 95(2): 214-21.

17. Pappalardo G, Frattaroli FM, Casciani E, et al. Retrorectal tumors: the choice of surgical approach based on a new classification. Am Surg 2009; 75(3): 240-8. 18. Lack EE, Glaun RS, Hefter LG, Seneca RP, Steigman C,

Athari F. Late occurrence of malignancy following resection of a histologically mature sacrococcygeal teratoma. Report of a case and literature review. Arch Pathol Lab Med 1993; 117(7): 724-8.

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