Thoracic wall tumor: A rare presentation of common disease

Niraj Kumar Srivastava[1], Sunita Singh[1], Nitin Kashyap[2], Vandita Singh[3], Debajyoti Mohanty[1]

From Departments of [1]Surgery, [2]Trauma and Emergency, and [3]Pathology, All India Institute of Medical Sciences, Raipur, Chhattisgarh, India

Correspondence to: Dr. Sunita Singh, Department of Surgery, All India Institute of Medical Sciences, Raipur - 492 099, Chhattisgarh, India. Phone: +91-8518887725. Tel,: 0771-2572733.

E-mail: drsunitasingh28@gmail.com

Received – 19 February 2017 Initial Review – 30 March 2017 Published Online – 20 May 2017

A 20-year-old female presented with a painless progressively increasing mass in the right hypochondrium of 3-month duration. On examination, an 8 cm × 3 cm (transverse, vertical dimension) parietal mass was palpable in the right hypochondrium. The lump was non-tender, firm, and having smooth surface. The lump became less prominent on Valsalva maneuver. The upper border was not reachable while the lower margin could be appreciated about 3 cm below the costal margin.

Contrast-enhanced computed tomography (CECT) abdomen, and lower thorax revealed a homogenous 8 cm × 7.3 cm × 3 cm mass arising from the anterolateral aspect of right lower 7th and 8th intercostal muscles in the mid-clavicular line. The tumor was showing minimal peripheral vascular enhancement (Fig. 1). The ultrasound-guided fine-needle aspiration cytology of the mass was inconclusive.

Complete excision of the mass was planned on the basis of CECT findings. Right subcostal incision was made. The mass was found to be arising beneath the right costal margin and bulging into peritoneal cavity (Fig. 2). Wide *en bloc* excision of the tumor with extraperiosteal segmental resection of ribs and diaphragm was carried out and sent for the histopathology (Fig. 3). The immunohistochemical (IHC) analysis was positive for Vimentin, smooth muscle actin, beta-catenin, and negative for S-100 P, CD-34, and membrane antigen. Ki-67 nuclear protein was positive <1%. Post-operative period was uneventful, and the patient discharged on the 6th post-operative day.

QUESTIONS

1. What can be the diagnosis?
2. What should be the next plan of management in this patient?

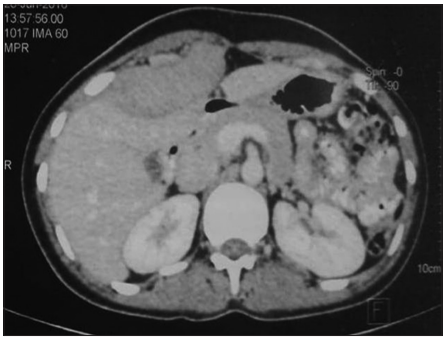

Figure 1: Contrast-enhanced computed tomography scan of abdomen axial view suggested a 8 cm × 7.3 cm × 3 cm (transverse, longitudinal and anteroposterior dimension) cm homogenous mass from the anterior abdominal wall extending beneath the right subcostal region. No clear plane was identified between mass and 3 lowest ribs anteriorly in mid clavicular line. The tumor showed mild peripheral vascular enhancement

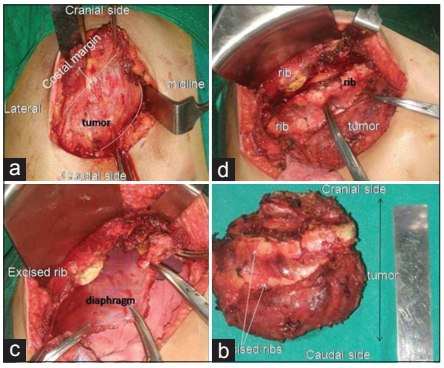

Figure 2: Peroperative photograph showing (a) mass protruding beneath the right costal margin (b) exposed ribs (c) after excision of mass tumor bed is shown with diaphragm margins, (d) excised sp ecimen with resected ribs

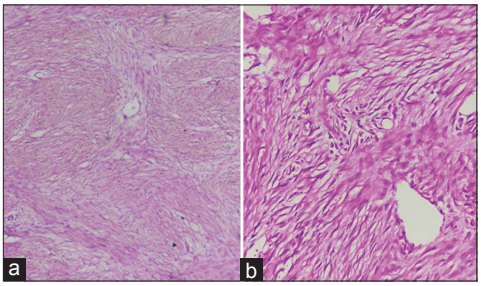

Figure 3: (a) Sweeping fascicles of spindle fibroblast separated by collagen within which are scattered thin walled ecstatic vessel, (H and E, ×40), (b) spindled fibroblasts and myofibroblasts with bland nuclei having tapered ends, very rare atypical mitosis, and foci of myxoid stromal change. Few perivascular lymphocytes and mast cells are present (H and E ×100)

ANSWER 1

The differential diagnosis of lower thoracic wall lump includes abdominal muscle hematoma, lipoma, liposarcoma, fibroma, fibrosarcoma, lymphoma, rhabdomyosarcoma, neurofibroma, and primitive neuroectodermal tumor. This was the case of desmoids tumor. The diagnosis was settled through histopathology and IHC analysis of the specimen.

Worldwide approximately 3.7 new cases of desmoids occur per 1 million persons per year [1]. The exact prevalence of desmoids tumor in India is still unknown. Desmoid tumors are also known as aggressive fibromatoses or low-grade fibrosarcoma [1,2]. Desmoids can occur in extremities (around limb girdles or proximal extremity), abdominal wall, in the bowel wall and mesentery. Patients with abdominal wall desmoids may have a prior history of abdominal surgery, trauma, pregnancy, estrogen therapy, familial adenomatous polyposis, and Gardner syndrome. The desmoids involving lower thoracic wall, and presenting as the right hypogastric lump is a rare entity.

Ultrasonography of desmoid tumor may show well-defined lesions with varying echogenicity [3]. The CECT appearance of desmoid may be hypo-, iso-, or hyper-intense (depending on their composition). The appearance may be homogeneous or heterogeneous as compared to attenuation of muscles [3]. Definitive diagnosis of desmoids can only be ascertained with histopathological examination [4].

Answer 2

Desmoid tumors are slow-growing benign tumors. Wide local excision is the first line of the management; however, these locally aggressive tumors have an increased potential for local recurrence despite adequate surgical excision [1]. The reported mortality rate from direct invasion of vital organs approaches 10%. Hence, periodic follow-up is mandatory in these patients. Recurrent tumors are preferably managed by re-excision of the tumor. Local adjuvant external beam radiation therapy in combination with chemotherapy can be offered to patients not consenting for revision wide local excision [5]. Pharmacological agents such as anti-estrogen therapy, cyclic-aminophylline (theophylline, chlorothiazide, ascorbic acid, and testolactone), warfarin, vitamin K-1, or prostaglandin inhibition (clinoril/sulindac) have been attempted with a variable degree of success [3].

REFERENCES

1. de Bree E, Keus R, Melissas J, Tsiftsis D, van Coevorden F. Desmoid tumors: Need for an individualized approach. Expert Rev Anticancer Ther. 2009;9(4):525-35.
2. Stoeckle E, Coindre JM, Longy M, Binh MB, Kantor G, Kind M, et al. A critical analysis of treatment strategies in desmoid tumours: A review of a series of 106 cases. Eur J Surg Oncol. 2009;35(2):129-34.
3. Bertagnolli MM, Morgan JA, Fletcher CD, Raut CP, Dileo P, Gill RR, et al. Multimodality treatment of mesenteric desmoid tumours. Eur J Cancer. 2008;44(16):2404-10.
4. Bhattacharya B, Dilworth HP, Iacobuzio-Donahue C, Ricci F, Weber K, Furlong MA, et al. Nuclear beta-catenin expression distinguishes deep fibromatosis from other benign and malignant fibroblastic and myofibroblastic lesions. Am J Surg Pathol. 2005;29(5):653-9.
5. Nuyttens JJ, Rust PF, Thomas CR Jr, Turrisi AT 3rd. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. Cancer. 2000;88(7):1517-23.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Srivastava NK, Singh S, Kashyap N, Singh V, Mohanty D. Thoracic wall tumor: A rare presentation of common disease. Indian J Case Reports. 2017;3(3):167-168.