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CASE REPORT

Desmoid-type fibromatosis arising in a bifid rib chest wall

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Abstract
Desmoid-type fibromatosis is a rare soft tissue tumor and the chest wall is one of the common sites of its extra-abdominal occurrence. A bifid rib is one of the congenital rib abnormalities. We report a case of desmoid-type fibromatosis arising in a chest wall's bifid rib. A 42-year-old female complained of right chest pain without remarkable medical, traumatic, or familial history. Chest-computed tomography revealed a chest wall tumor located adjacent to a bifid costal cartilage of third rib. We performed chest wall resection of second and third ribs. Pathologically, the tumor was diagnosed a desmoid-type fibromatosis of the chest wall. We surmise mechanical stimulation due to the bifid rib may be related to the occurrence of the tumor. In case of desmoid-type fibromatosis without somatic gene mutation, traumatic history, wound, implants, or use of female hormonal agents, we should search also local congenital abnormality.

Keywords Desmoid · Fibromatosis · Chest wall · Bifid rib

Introduction

Desmoid-type fibromatosis is a rare mesenchymal tumor with an incidence of 2.4–4.3 cases per 1,000,000 per year which locally infiltrates, but does not metastasize [1, 2]. Chest wall is one of the common sites of extra-abdominal desmoid-type fibromatosis [3].

Bifid rib is a congenital rib abnormality with a prevalence that ranges between 0.15 and 3.4% [4]. Most bifid ribs are usually asymptomatic and found incidentally [5].

We report a rare case of desmoid-type fibromatosis arising in the chest wall's bifid rib.

Case

A 42-year-old non-smoking female complained of a neuralgia-like right chest pain. The patient had no remarkable medical, traumatic, or familial history. Chest-computed tomography revealed a chest wall tumor from the second rib to the third rib. The tumor grew mainly into the intrathoracic space and partially into the extra-chest wall region through the second intercostal space. Computed tomography also revealed bifurcation of the costal cartilage of the third rib resulting in a "bifid rib" with the center of the tumor located adjacent to the cranial side of the bifid rib (Fig. 1). We performed chest wall resection of the second and third ribs followed by reconstruction with a polytetrafluoroethylene mesh. Operative time was 183 min and blood loss during surgery was 90 ml. A defect of chest wall was 15 × 10 cm. Margin distances of cranial side, caudal side, sternal side, and vertebral body side from the tumor edge were 1 cm, 1.5 cm, 1 cm, and 1 cm, respectively. The patient had her chest tube removed at postoperative day 2 and was discharged at postoperative day 8 without any complication. Her neuralgia-like chest pain decreased gradually and disappeared within postoperative 2 months.

Gross view of the surgically resected specimen showed a white mass arising from the cranial side of the bifid costal cartilage of the third rib (Fig. 2). Microscopic examination revealed spindle tumor cells without nuclear atypia proliferating with fibromatosis (Fig. 3a). Immunohistochemistry revealed that the tumor cells were positive for β-catenin (Fig. 3b) and Vimentin and negative for CD34, S100, and C-KIT. Based on these findings, the tumor was diagnosed as a desmoid-type fibromatosis. Pathologically, there was no

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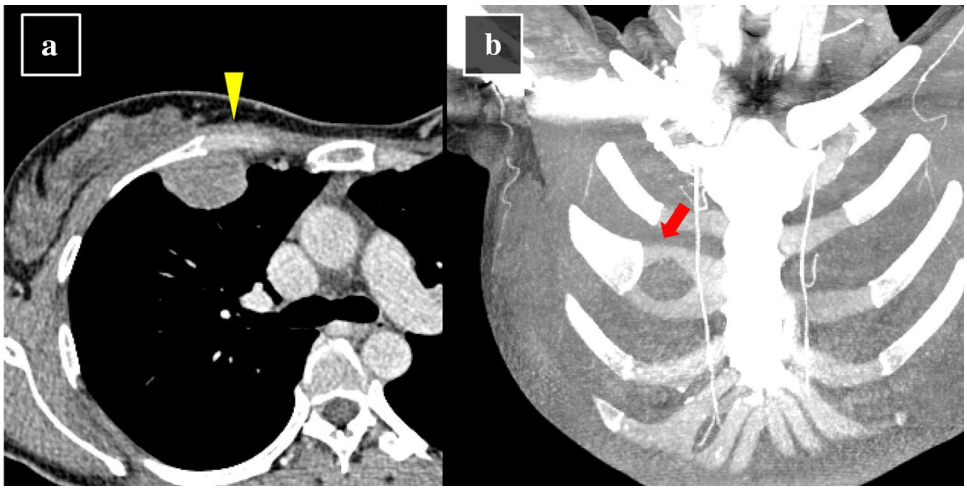

Fig. 1 **a** Chest wall tumor grew into intrathoracic space and center of the tumor located adjacent to the cranial side of the bifid third costal cartilage (arrow head). **b** Costal cartilage of third rib was bifid rib (arrow)

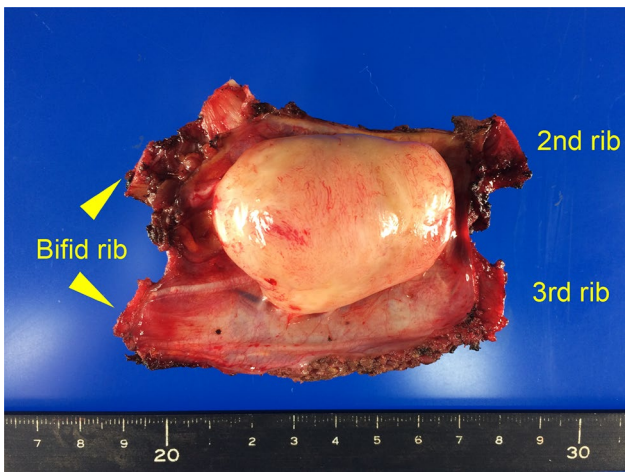

Fig. 2 Gross view from intrathoracic space side showed a white mass arising from the location of cranial side of bifid fib

evidence to suggest a relationship between the occurrence of the tumor and the bifid rib.

In addition, there was no somatic CTNNB1 gene mutation. The patient was not placed on medications for recurrence prevention such as COX-2 inhibitor or hormone therapy. Presently, there is no evidence of recurrence at 30 months after surgery.

Discussion

In this case, we suspected the presence of the bifid rib was related to the occurrence of desmoid-type fibromatosis.

Somatic gene mutation of APC or CTNNB1 is known to be related to oncogenesis of desmoid-type fibromatosis [1–3]. Conversely, the etiology of desmoid-type fibromatosis without genetic mutations has not been sufficiently elucidated. Traumatic history, wound scars, implants of the breast, and female hormones have been reported as factors

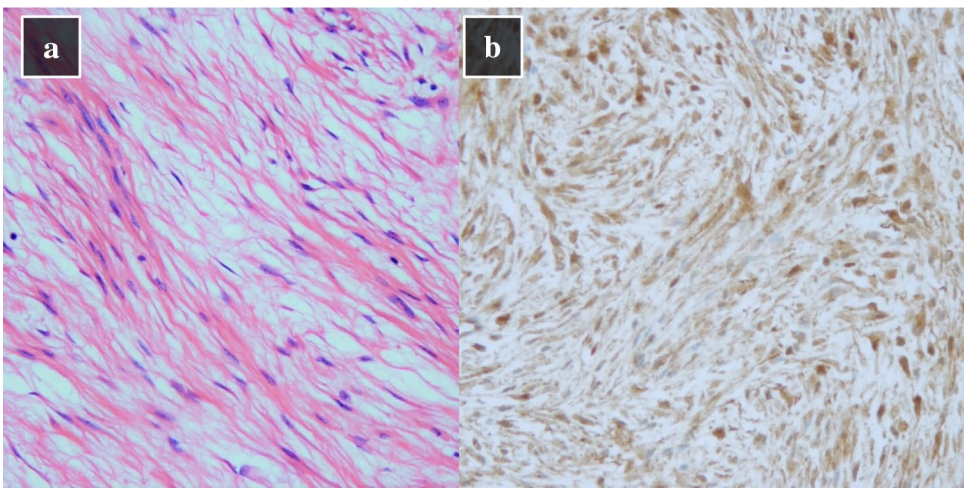

Fig. 3 **a** Spindle tumor cells without nuclear atypia proliferated accompanying by fibromatosis. **b** Tumor cells were positive for β-catenin

related to the occurrence of desmoid-type fibromatosis [2]. The patient had no mutation of CTNNB1, traumatic history, colorectal polyp, or notable familial history. Although the relationship could not be pathologically proven in this case, we surmise that mechanical stimulation due to the bifid rib may be related to occurrence of the tumor.

We searched databases of Pubmed, Ovid MEDLINE, and The Cochrane Library with the words "desmoid" and "bifid rib" and found no article. Therefore, we considered this case with the simultaneity of these two rare diseases to be valuable as a hint for elucidation of etiology of desmoid-type fibromatosis without genetic mutations.

Conclusion

We surmised that mechanical stimulation due to the bifid rib may be related to occurrence of the desmoid-type fibromatosis. In case of desmoid-type fibromatosis without somatic gene mutation, traumatic history, wound, implants, or use of female hormonal agents, we should search also local congenital abnormality.

Compliance with ethical standards

Conflict of interest The authors have declared that no conflicts of interest exist.

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