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Case Report

STRATOS Titanium Rib Bridge for Chest Wall Reconstruction after Infantile Fibrosarcoma Resection: A Case Report

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Resection and reconstruction of the chest wall for the radical surgical treatment of malignant soft tissue tumors are currently considered a substantial challenge for thoracic surgeons. We present an unusual case of infantile fibrosarcoma with tropomyosin 3-neurotrophic receptor tyrosine kinase 1 fusion in a 13-year-old patient. The surgical treatment consisted of radical resection of the right posterior chest wall and reconstruction with the use of the STRATOS (Strasbourg Thoracic Osteosynthesis System) titanium rib bridge system. The patient had a favorable postoperative course and received respiratory-ventilatory rehabilitation, adjuvant therapy with chemotherapeutic agents, immunotherapy, and radiotherapy.

Keywords: Infantile fibrosarcoma, Chest wall resection and reconstruction, STRATOS, Titanium plate, Rib prosthesis, Case report

Case report

Infantile fibrosarcoma (IF) is a rare entity that originates from primitive mesenchymal cells and represents fewer than 1% of childhood malignant tumors [1]. It mainly affects the extremities (73%) and rarely the thorax (1.8%) in infants under 1 year of age [2]. The main treatment is radical surgical resection; however, when it affects the thoracic region, resection is usually accompanied by reconstruction with various osteosynthesis techniques or devices. The resection is usually extensive and is rarely well-tolerated, leading to high morbidity due to biomechanical alterations of the chest wall. This has led thoracic surgeons to consider using osteosynthesis systems such as Strasbourg Thoracic Osteosynthesis System (STRATOS; MedXpert GmbH, Heitersheim, Germany) to provide mechanical stability and an "anatomically equivalent" chest wall with better physiological results [3].

A 13-year-old male adolescent with no relevant history attended an outpatient consultation due to progressively presenting dyspnea and mild chest pain in the dorsal infrascapular region associated with a progressively growing tumor for 6 years. Chest auscultation revealed no abnormal findings, and palpation revealed a tumor measuring 11×9 cm with an indurated, infiltrative consistency and without defined borders.

Thoracic radiography showed an inferior medial right radiopacity, and perfusion tomography showed a tumor in the right posterior thoracic wall (latissimus dorsi muscle) that extended from the seventh to the tenth costal arch, without intrathoracic involvement and close to the transverse processes of the thoracic vertebrae (Fig. 1). Percutaneous biopsy, immunohistochemistry, and genomic studies revealed a non-metastatic IF with tropomyosin 3 (TPM3)–neurotrophic receptor tyrosine kinase 1 (NTRK1) fusion and intron 7 rearrangement, with therapeutic susceptibility to larotrectinib and entrectinib.

Radical surgical resection of the IF and reconstruction of the chest wall with the STRATOS titanium rib bridge system were indicated. The tumor was excised with the surrounding tissue and parietal pleural tissue, with partial resection of the seventh to the ninth costal arches and total disarticulation of the 10th costal arch. The proximal areas of the costovertebral (4 cm) and chondrocostal (13 cm) joints of the partially resected rib arches were preserved and later used as anchoring areas for the thoracic osteosynthesis system. The space created by the resection was replaced with the implantation of a polypropylene mesh,

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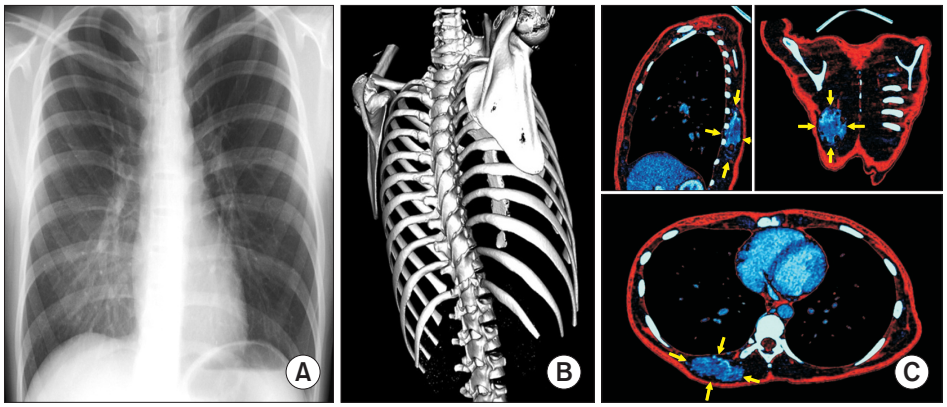

Fig. 1. Preoperative imaging studies. (A) Chest X-ray showed an inferior medial right radiopacity. (B) Computed tomographic study with 3-dimensional bone reconstruction of the thorax. (C) Tomography-perfusion study showing an infiltrating tumor of the right posterior chest wall (arrows).

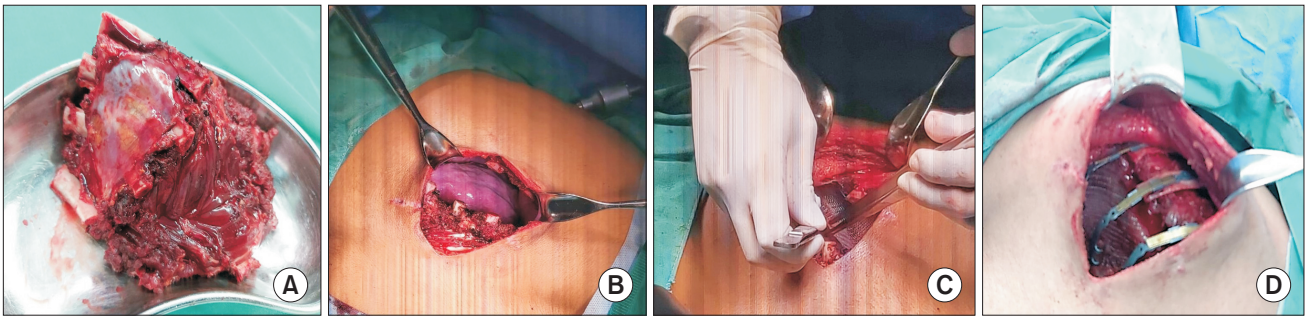

Fig. 2. Surgical procedure of chest wall reconstruction. (A) Surgical specimen of the radical tumor resection of the posterior chest wall, which also contains surrounding tissue, costal arches, and parietal pleura. (B) The surgical space created by thoracic resection. (C) Placement of polypropylene mesh as parietal pleura and fixation to the residual rib ends. (D) Implantation of the STRATOS (Strasbourg Thoracic Osteosynthesis System) titanium rib bridge system from the seventh to ninth rib arches. Written informed consent for publication of this image was obtained from the patient.

simulating the parietal pleura and adhering to the residual rib ends as a thoracic protection and reinforcement mechanism. Three STRATOS titanium rib bridge systems were placed from the seventh to the ninth rib arches, anchored in the previously described rib segments, and osteosynthesis of the tenth rib arch was not performed due to its previous total resection and the floating anatomical configuration. Each bridge consisted of 2 clips and 1 connecting bar crimped to each other and the rib (Fig. 2).

The operation was uneventful, and the postoperative stay was short and without complications. The patient was discharged from the hospital with an indication for respiratory-ventilatory rehabilitation, adjuvant therapy 30 days after surgery, and periodic outpatient pediatric examinations to assess possible future repercussions of STRATOS and the growth and development of the patient. The adjuvant therapeutic regimen consisted of chemotherapeutic agents, radiotherapy, larotrectinib, and entrectinib. Eighteen months after surgery, the patient is disease-free, without recurrence or metastasis (Fig. 3).

This case report was evaluated and approved by the Edgardo Rebagliati Martins National Hospital Ethics Committee (RCEI-7/HNERM/0076-09). Written informed consent for publication of this image was obtained from the patient.

Discussion

Chest wall resection and reconstruction due to the radical surgical treatment of malignant soft tissue tumors are currently considered a substantial challenge for thoracic surgeons. Furthermore, facing an unusual case of IF in a non-pediatric patient is of particular interest and requires thoroughness in the curative surgical strategy. The fundamental principle of oncological thoracic resection is to guarantee disease-free margins; however, it is accompanied by considerable alterations in respiratory dynamics due to the instability it generates and the imminent risk of pulmonary hernia and associated intrathoracic infections [3]. Reconstruction of the chest wall becomes necessary and is

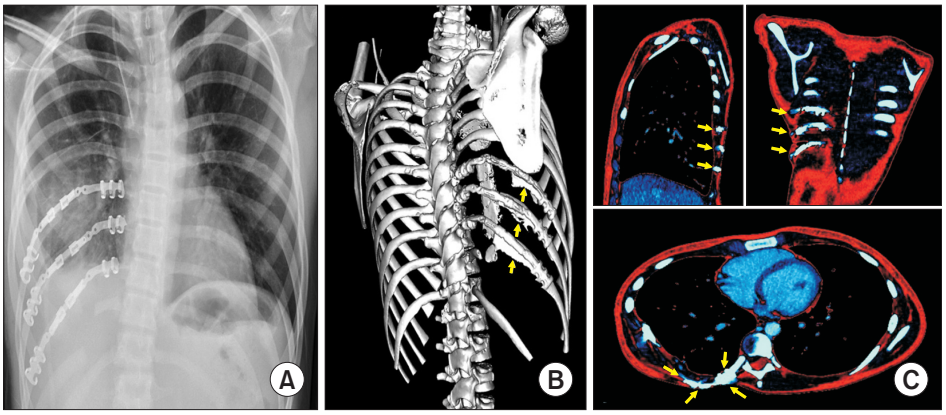

Fig. 3. Postoperative imaging studies. (A) Chest radiograph showing the rib osteosynthesis system. (B) Computed tomographic study with 3-dimensional bone reconstruction of the thorax, which shows the correct implantation of the 3 STRATOS (Strasbourg Thoracic Osteosynthesis System) titanium rib bridge systems (arrows). (C) Tomography-perfusion study showing absence of any residual tumor and visualization of the thoracic osteosynthesis system (arrows).

indicated when the resection exceeds 5 cm in the anterior region and 10 cm in posterior areas close to the scapula; the aim of reconstruction is to restore the integrity of the wall, preserve lung compliance, and maintain the impermeability of the pleura [4].

The choice of material and the thoracic reconstruction strategy are essential to achieve optimal functional, aesthetic, and postoperative survival results. It has been proposed that the ideal characteristics of a prosthetic material are stiffness, to inhibit the paradoxical movement of the chest wall; inertia, to allow the growth of the fibroelastic tissue and to maintain the aesthetic properties of the reconstruction; and radiolucency, to enable easy radiographic follow-up after implantation [5]. Titanium is an invaluable therapeutic option due to these characteristics, together with its high resistance to body weight, corrosion, and infection, as well as its properties in terms of osseointegration and non-ferromagnetism.

As has been described in multiple studies, titanium plates yield superior results for thoracic reconstruction because these adapt to the thoracic curvature and maintain their shape and volume. There are currently 2 titanium osteosynthesis systems on the market, STRATOS and the Matrix Rib (DePuy Synthes, Raynham, MA, USA) [4]. The first, as previously described, uses staples with screws that are attached to the end of each rib, which is attached with titanium bars. In the Matrix Rib system, the plate is attached directly to the rib with screws; the main advantage of this system is that it does not require resecting the periosteum and allows maximum vascular conservation. Despite the aforementioned advantages, Edgardo Rebagliati Martins National Hospital only has STRATOS, which was the option chosen for the case described herein; however, we are aware of the existence of other osteosynthesis systems for post-resection thoracic reconstruction [1].

The STRATOS titanium rib bridge system has frequently been used in thoracic trauma and has recently been considered as an option in thoracic reconstruction due to tumor resection. Its use has been described in 61% of chondrosarcomas, 20% of osteosarcomas, and 6% of Ewing sarcomas, while no evidence has been published regarding IF of the rib wall [4]. In a study of patients who received reconstruction with STRATOS, 96% of patients were extubated early during the first postoperative day, patients had shorter stays in the intensive care unit, and no mortality was observed in the first 30 postoperative days thanks to a rapid recovery of lung function [3,4,6]. Likewise, a 3%–10% local recurrence rate has been reported at 5 years after thoracic resection and reconstruction, and the reported overall survival rates at 5 and 10 years are 60%–80% and 45%–60%, respectively; however, these data do not include cases of IF [3-5].

The report of an IF in the chest wall with TPM3-NTRK1 fusion in an adolescent patient, in whom the STRATOS system was used for reconstructive therapy after excision, is extremely rare in the medical literature; currently, there are no follow-up reports or survival data for this category of patient. However, some studies of resected IF in pediatric patients without thoracic involvement, which could provide insights into this unusual condition, have reported an overall survival of 84%–93% at 5 years after resection [1]. Furthermore, we also would like to highlight the importance of TPM3-NTRK1 fusion in our case, because this is an extremely rare genomic mutation (<1%) that triggers a variety of little-known and insufficiently-studied soft tissue neoplasms, such as neural lipofibromatosis-like tumor, uterine and vaginal sarcoma, malignant peripheral nerve sheath tumor-like, myofibroblastic sarcoma-like tumor, myopericytoma-like tumor, IF and more [7,8].

We believe that this unique case confirms the impor-

tance of chest wall reconstruction after extensive cancer resection, allowing rapid postoperative recovery and restoration of respiratory dynamics. The STRATOS system is an excellent and user-friendly surgical strategy that offers countless options for thoracic reconstruction, and its use requires adequate training among today's thoracic surgeons. There is an urgent need for further studies that would allow us to understand the behavior and development of soft tissue tumors with unusual genomic and immunohistochemical characteristics, and the findings of such studies would make it possible to implement therapeutic regimens based on surgical resection and appropriate adjuvant therapies.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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