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Surgical treatment of retrosternal extraosseous Ewing Sarcoma in a 6-years old female: A clamshell approach with hemysternectomy and application of a non-crosslinked extracellular matrix

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ABSTRACT

Background: Ewing Sarcoma (ES) and Neuroblastoma (NB) belong to a family of tumours of primitive neuroectodermal origin (PNET) that occurs in both bone and soft tissue. Notwithstanding ES and NB are two distinct malignant tumours, sometimes there could be a link between them.
Case report: We describe a case of an extraosseous ES localized in the retrosternal region and the upper lobe of the right lung, which had been previously treated for NB in a 6 years old female. We treated this case with a clamshell approach which allows, in a one-step surgery, a complete excision of the mass reconstructing the hemysternectomy with a non-crosslinked matrix.
Conclusion: the clamshell approach is therefore useful to achieve the retrosternal space and the lung with a single surgical access. According to our experience, we consider appropriate to use a non-crosslinked matrix for sternal reconstruction.

1. Introduction

Ewing Sarcoma (ES) and Neuroblastoma (NB) belong to a family of tumours of primitive neuroectodermal origin (PNET) that occurs in both bone and soft tissue. The PNET/ES are small round neoplasms cells without cellular or structural differentiation [1] ES is the second most commonly malignant bone tumour in the childhood [2]. NB occurs typically in the early childhood and is localized on the sympathetic ganglia and in the adrenal medulla [3]. Notwithstanding ES and NB are two distinct malignant tumours [4], and sometimes there could be a link between them [5].

While for NB the treatment is a surgical removal of the mass, the ES requires a combination of chemotherapy, before or after surgery, and, if necessary, radiotherapy [5] [6].

In our case report we show a 6-years old girl with an extraosseous ES localized in the retrosternal region and the upper lobe of the right lung, which had been previously treated for NB. Due to the unusual localization we have used a clamshell approach which allows, in a one-step surgery, the complete excision of the mass reconstructing the hemysternectomy with a non-crosslinked extracellular matrix.

2. Case report

A 6-years-old girl was admitted to our Department with the diagnosis of extraosseous Ewing sarcoma of the anterior mediastinum and the upper lobe of the right lung.

Two years earlier, the patient had been hospitalized at another hospital for abdominal pain. A chest X-ray had showed a right thoracic mass with pleural effusion; a total body CT scan had showed a mass of 12 × 12 × 7.6 cm, adherent to pericardium, mediastinal and diaphragmatic pleura and liver dome. A bone marrow aspirate was negative; a needle biopsy reported the presence of neuroblastoma; MIBG scintigraphy confirmed the presence of a mass with enhanced uptake.

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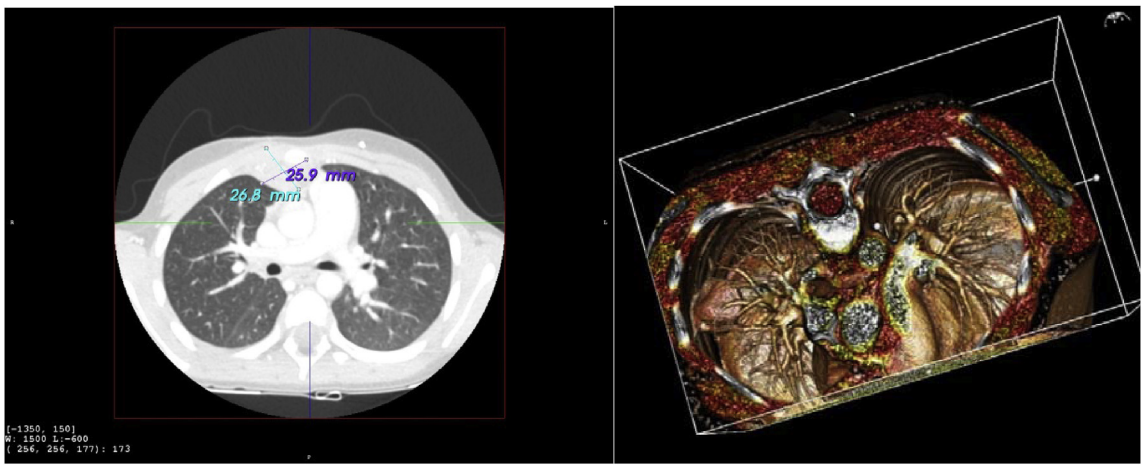

Fig. 1. CT-scan and 3D reconstruction of the retrosternal and upper lobal mass.

After two cycles of carboplatin/VP and two cycles of CADO chemotherapy (according to the "European Low and Intermediate Risk Neuroblastoma Protocol: A Siopen Study"), a new chest and abdomen CT scan showed a reduction of the mass.

Few months later the girl was subjected to removal of the residual mass, chemotherapy (Carboplatin/VP16 and CADO) and radiotherapy.

Following occurrence of cough and shortness of breath, a CT

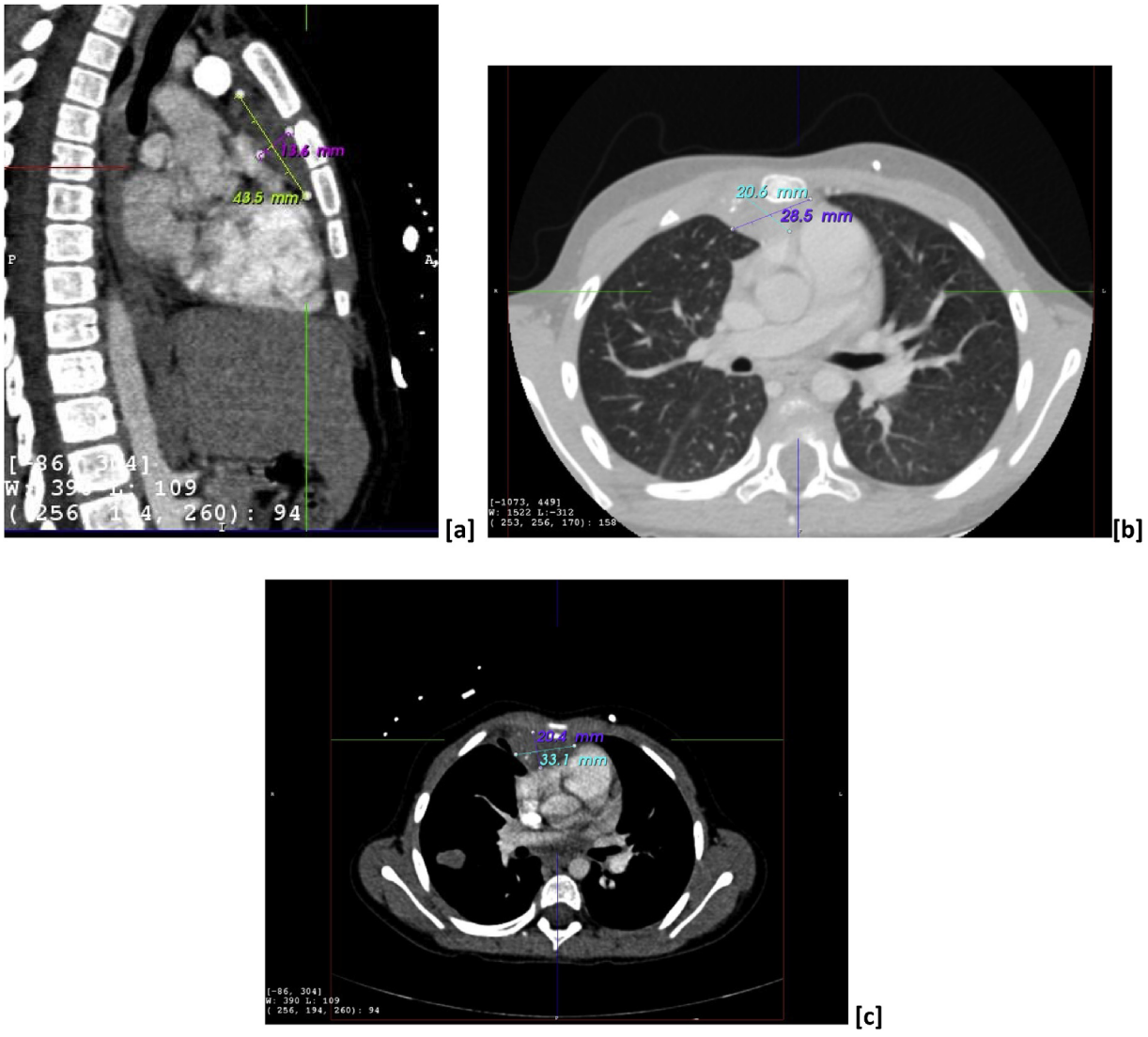

Fig. 2. [a-b-c]: Preoperative CT scan shows the reduction of the retrosternal mass after the "Eudra CT:2008 – ISG AIEOP EW1" protocol. [a] axial; [b-c] coronal.

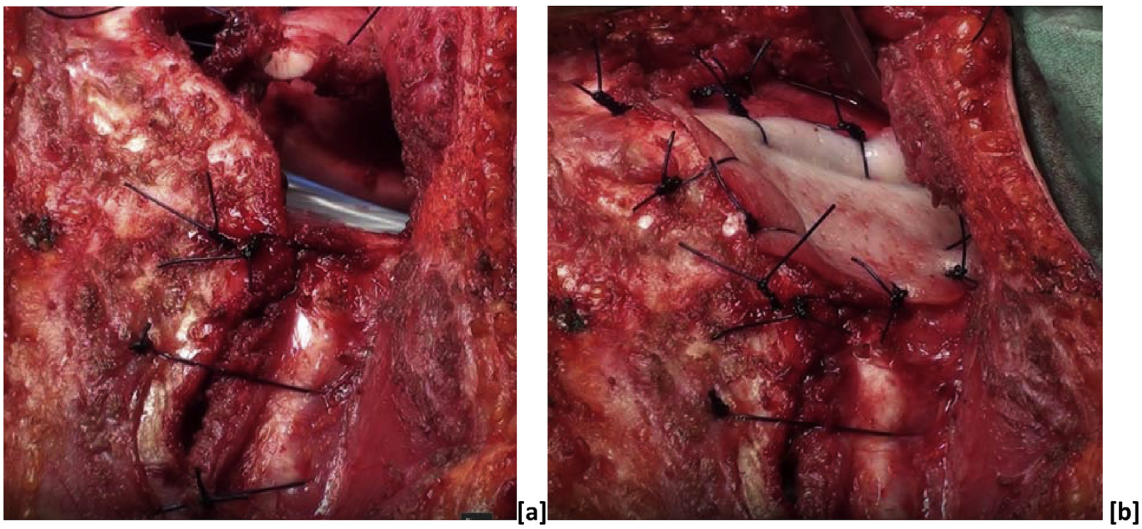

Fig. 3. [a-b]: Hemysternectomy [a]; the sternum was reconstructed using a non-crosslinked matrix [b].

scan showed a mass of 33 × 23 × 18 mm in the upper lobe of the right lung, pleural nodules of 5 mm and multiple lymphadenopathy, and a mediastinal mass of 42 × 35 × 43 [Fig. 1]. MIBG scintigraphy showed increased uptake of the drug and no bone uptake; bone marrow aspirate was negative. Mediastinal biopsy and histologic examination showed the presence of Ewing sarcoma, with positive molecular analysis for the detection of EWS/FLI1 t(11; 22) (q24/12).

Treatment was then started according to the "Eudra CT:2008 – ISG AIEOP EW1" protocol. A new chest CT scan showed the presence of a 2,6 × 2,2 × 1,6 cm mass in the upper lobe of the right lung, a 5 mm pleural nodule, a mediastinal mass of 3,7 × 2,6 × 5 cm, an infiltration of the chondrosternal cartilage and soft tissue on the right parasternal zone and the reduction of lymphadenopathy.

When the patient came to our observation, a chest CT scan was performed, reporting a reduction in the volume of the mass at the upper lobe of the right lung, and no signs of infiltration of the sternum and the chondrosternal cartilage **[**Fig. 2 **a-b-c]**.

The child was then subjected to surgical removal of the mediastinal mass, of the right sternum (due to the positive result of the extemporaneous histological examination) and two costal cartilages, after clamshell incision (double thoracotomy with longitudinal sternotomy). Then the bulk of the upper lobe of the right lung was found and removed; the sternum was reconstructed using a non-crosslinked matrix (XenMatrix®) [Fig. 3 **a-b**, Fig. 4]. Histological examination showed the presence of extraosseus Ewing sarcoma/pNET. The postoperative recovery was uneventful.

After four months, before chemotherapy, a re-staging CT and a Pet-scan showed a mass of 2 cm in the basal right lobe, with suspected residual/recurrent disease [Fig. 5]. The child was then subjected to high-dose chemotherapy and then autologous stem cells transplantation and administration of GCSF. She was discharged in good general condition.

The patient was hospitalized again at our Department. We performed a surgical removal of the right diaphragmatic metastases of extraosseous Ewing's sarcoma. Through a right posterolateral transpleural access, a central portion of the right diaphragm was resected with removal of metastases and the diaphragm was closed by a primary closure with nonabsorbable suture. Then, an atypical resection of a portion of the right lower lung lobe was performed. Histological examination showed the presence of an extraosseus Ewing sarcoma/pNET.

Three month after the surgery, a CT scan didn't show any residual disease nor morphological alteration of the chest wall.

3. Discussion

This case report describes a surgical treatment of an EOE relapse after a previous surgical excision of a thoracic NB. The group of Ewing Sarcoma comprises both osseous and extra osseous tumours. There is a large consensus about considering PNET/ES, NB and EOE as the same entity showing varying degrees of neuroectodermal differentiation, and the differences can be sometimes overlapping [5,7]. NB is originated from primordial neural crest cells while the histogenesis of PNET/ES is still controversial: a recent hypothesis

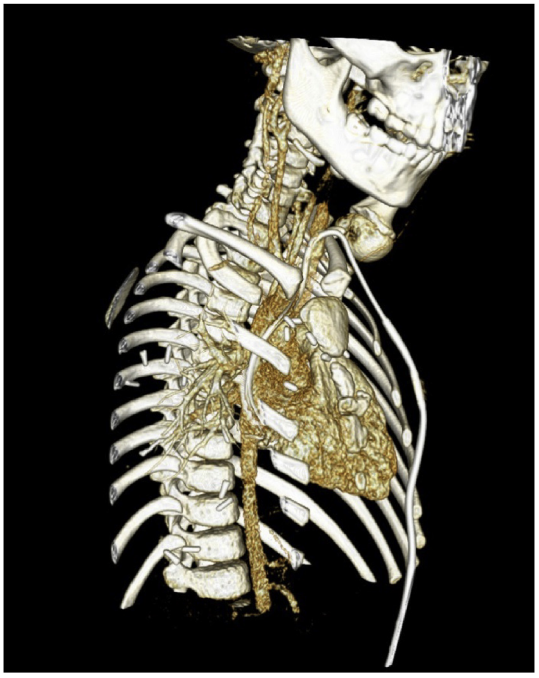

Fig. 4. 3D reconstruction (from CT-scan) after four month from the surgery shows the bony callus related to the non-crossed linked matrix.

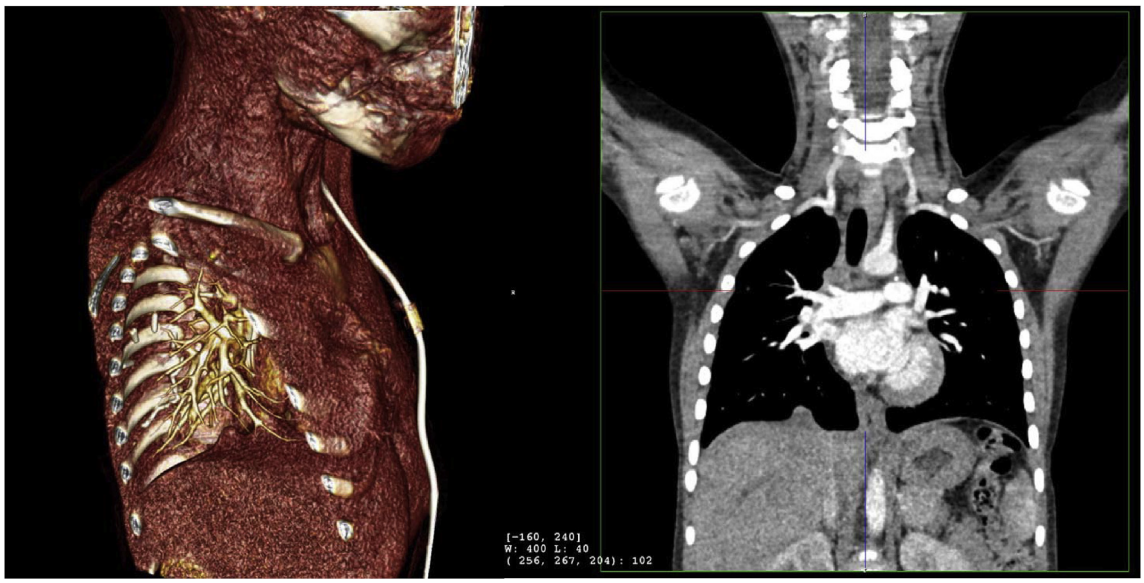

Fig. 5. 3D reconstruction (from CT-scan) after four month from the surgery. It is shown the diaphragmatic mass.

considers that it might arise from mesenchymal stem cells [5].

EOE arises most commonly from the paravertebral region, but it is also reported that EOE involves the soft tissue of the orbit and vagina, as well as kidney and posterior mediastinum [6]. It usually manifested as an individual soft tissue mass, painless but occasionally with local symptoms, growing rapidly with distant metastases at the time of diagnosis approximately in 25% of patients [7].

We can considered the stage and the subcutaneous lesions as a prognostic factor for patients affected by EOE, whereas other factors as margin status, site, tumour size and the presence of residual tumour after neoadjuvant therapy can't be reasonably included among the predictive factors of survival [2]. In addition to that, the relapse is another important negative prognostic factor, in particular related to its time (<o > 2 years), type (local, systemic or combined relapse) and site (a lung relapse is better than in another site) [5].

The therapy of EOE now includes a combined chemotherapy, surgery and, sometimes, radiotherapy but it is less effective than in ES [8]. Some studies have shown that there is no difference between the adoption of chemotherapy before or after surgery in patients with localized disease. On the other hand, in the metastatic disease or in unresectable recurrent disease only chemotherapy is utilised to improve the progression-free survival [9].

The survival of EOE is around 61–65% but it is different between localized and metastatic/recurrent diseases; the patients with localized disease have a 5-years survival rate of 70% with a relapse in approximately 30% of patients, while for the metastatic/recurrent the rate is around 25%.

Do to the retrosternal and upper right lobe localisation of ES, we have choose to trait our patient with a surgical resection of the mass through a clamshell approach. This is an approach more utilised in the adult surgery than in paediatric age, where his use is limited to cardiac surgery, lung transplantation and the resection of lung metastasis [10–12].

The clamshell approach presents some advantages for this kind of surgery because in a one-stage surgery we can explore all the lungs without changing the patients' position [13,14]. Moreover, this approach has a low rate of operatory complications and it have an optimal tolerance from the patients [11]. On the other hand, this approach, in addition to those linked to the possible complications of the surgery, where the most important is the sternal dehiscence that have an incidence rate of 8–36% [15], shows some disadvantages linked to the surgical technique. These last problems, due to the long-life expectation, are important because this approach could cause a preclusion of breast development in females and it precludes a possible future use of both internal thoracic arteries for a coronary grafting [11]. Due to the not perfect vision of the diaphragm that the clamshell approach permits for the diaphragmatic relapse, a different approach it has been adopted during the second surgery.

Although the stainless wires, both sternal uncrossed and parasternal crossed are widely used for the reconstruction of the chest wall [11,12,14–16], but there are many different techniques. Saenz et al. in their retrospective study of 20 cases of chest wall Ewing's Sarcoma-primitive neuroectodermal tumour suggest, as alternative possibility to the Gore-Tex, a reconstruction with a synthetic Marlex mesh that can permit to achieve adequate margin and have an acceptable functional and cosmetic result [17]. Orsini et al. in their study of three cases propose, both as preventive technique and as immobilization of a sternal dehiscence after a clamshell incision, the indirect stabilization with the Stratos device, a titanium device that can reduce the risk of infection and must be fixed to the ribs instead of the sternum [18]. We have decided to utilise the Xenmatrix® cause to the cell regeneration allows by the matrix and for the strength required for the reconstruction of the hemysternectomy [19].

In conclusion, the clamshell approach is therefore useful to achieve the retrosternal space and the lung lobe with a single surgical access. According to our experience, we consider appropriate to use a non-crosslinked matrix for sternal reconstruction.

References

[1] Paoletti H, Colineau, Acalet L, et al. Ewing's sarcoma of the soft tisues: apropos of 3 cases and review of the literature. s.l. J Radiol 1999;80:477–82.
[2] Shannon Orr W, Denbo Jason W, Billups Catherine A, Jianrong WU, Navid Fariba, Rao Bhaskar N, et al. Analysis of prognostic factors in extra-osseous ewing sarcoma family of tumor: review of St. Jude Children's research hospital experience. s.l. Ann Surg Oncol 2012;19:3816–22.
[3] Messina M, Di Maggio G, Garzi A, Molinaro F, Amato G, Ferrara F. Neonatal neuroblastoma and prenatal diagnosis. Minerva Pediatr 2009;61(3):349–51.
[4] Weiss SW, Goldblum JR. Ewing's sarcoma/PNET tumor family and related lesions. 2008. p. 945–88.

[5] Vali K, Kokta V, Beaunoyer M, Fetni R, Teira P, Ssartelet H. Extraosseous Ewing sarcoma with foci of neuroblastoma-like differentiation associated with EWSR1 (Ewing sarcoma breakpoint region 1)/FLI1 translocation without prior chemotherapy. s.l. Hum Pathol 2012;43:1772–6.
[6] Stahl M, Ranft A, Paulussen M, Bolling T, Vieth V, Bielack S, et al. Risk of recurrence and survival after relapse in patients with Ewing sarcoma, vol. 57; 2011. p. 549–53.
[7] Siddiqui MA, Akhtar J, Shameen M, Baneen U, Zaheer S, Shahid M. Giant extraosseous Ewing sarcoma of the lung in a young adolescent female - a case report. s.l. Acta Orthop belg 2011;77:270–3.
[8] Krasin MJ, Davidoff AM, Rodriguez-Galindo C, Billups CA, Fuller CE, Neel MD, et al. Definitive surgery and multiagent systemic therapy for patients with localized Ewin sarcoma family of tumors. Local outcome and prognostic factors. Am Cancer Soc 2005:367–73.
[9] Galyfos G, Karantzikos GA, Kavouras N, Sianou A, palogos K, Filis K. Extraosseous ewing sarcoma: diagnosis, prognosis and optimal management. Indian J Surg 2016;78:49–53.
[10] Yutaka H, Hiromasa Y, Koichi I, Shinichi T, Shinichiro M. Surgical resection of a massive primary mediastinal liposarcoma using clamshell incision combined with lower median sternotomy: report of a case. Ann Thorac Cardiovasc Surg 2014:606–8.
[11] luciani GB, Starnes VA. The clamshell approach for the surgical treatment of complex cardiopulmonary pathology in infants and children. Eur J Cardiothoracic Surg 1997;11:298–306.
[12] Luciani GB, Wells WJ, Khong A, Starnes VA. The clamshell incision for bilateral pulmonary artery reconstruction in tetralogy of Fallot with pulmonary atresia. J Thorac Cardiovasc Surg 1997:443–52.
[13] Takuro M, Naoya Y, Tomoshi T, Keitaro M, koichi T, Kuniko A, et al. Left sleeve pneumonectomy via a clamshell incision for lung cancer with carinal invasion: report of a case. Surg today 2012;42:593–6.
[14] Abbo O, Guatta R, Pinnagoda K, Joseph J-M. Bilateral anterior sternothoracotomy (clamshell incision): a suitable alternative for bilateral lung sarcoma metastasis in children. world J Surg Oncol 2014;12:233–7.
[15] Costa J, Sonett JR, D'Ovidio F. Modified transverse thoracosternotomy and cost-effective reinforced sternal closure. Ann Thorac Surg 2015;100:2376–8.
[16] Miyazaki T, Yamasaki N, Tsuchiya T, Matsumoto K, Tomoshige K, Abe K, et al. Left sleeve pneumonectomy via a clamshell incision for lung cancer with carinal invasion: report of a case. Surg today 2012;42:593–6.
[17] Saenz NC, Hass DJ, Meyers P, Wollner N, Gollamudi S, Bains M, et al. Pediatric chest wall Ewing's Sarcoma. J Pediatr Surg 2000;35:550–5.
[18] Orsini B, D'Journo XB, Reynauld-aubert M, Thomas PA. Sternal dehiscence after clamshell incision in lung transplantation treated with the STRAsbourg Thoracic Osteosyntheses System. Ann Thorac Surg 2014;97:e55–7.
[19] Molinaro F, Garzi A, Cerchia E, Di Crescenzo V, Luzzi L, Bullotta AL, et al. Sternal reconstruction by extracellular matrix: a rare case of chaces syndrome. Open Med 2016;11:196–9.