

Psychosocial rehabilitation of a young patient through interscapular thoracic disarticulation and hemithoracectomy

Reinserção psicossocial de paciente jovem através de desarticulação interescápulo torácica e hemitoracectomia

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ABSTRACT

In the era of minimally invasive surgery, advocating large operations seems controversial. However, there is still room for major oncologic interventions. This is report of a 23 year-old patient with desmoid tumor in the right hemithorax, deformed and eroded ribs, ipsilateral lung partial atelectasis, contralateral mediastinal compression and shift, invasion of nerves in the brachial plexus, subclavian vessels and right apical pleura. Thoracotomy was performed with resection of the tumor in the mediastinum, right upper limb amputation, scapulectomy, and chest wall reconstruction with methylmethacrylate and marlex screen, and use of a fasciocutaneous flap. The subject is in follow up as an outpatient and has almost recovered his daily functions, which are now limited only for activities involving the inarticulate member. Selected cases should be considered for major interventions with aiming at R0 resection (resection with free surgical margins) and improved quality of life, always bearing in mind the need for rehabilitation and socialization.

Key words: Fibromatosis, Aggressive; Thoracoplasty; Disarticulation; Rehabilitation; Socialization.

RESUMO

Na era das cirurgias minimamente invasivas, parece controverso advogar grandes operações. Entretanto, ainda há espaço para grandes intervenções oncológicas. É relatado caso de paciente de 23 anos de idade portador de tumor desmoide em hemitórax direito, com deformidade e erosão de arcos costais, atelectasia parcial do pulmão ipsilateral, compressão e desvio contralateral do mediastino, invasão de nervos do plexo braquial, vasos subclávios e pleura apical direita. Realizada toracectomia, ressecção de tumor em mediastino, amputação do membro superior direito, escapulectomia e reconstrução da parede torácica com tela de márlex e metilmetacrilato e confecção de retalho fasciocutâneo. O paciente encontra-se em seguimento ambulatorial com suas funções diárias quase recuperadas, limitado apenas nas atividades que envolviam o membro desarticulado. Em casos selecionados devem ser consideradas grandes intervenções com o objetivo de ressecção R0 (ressecção com margens cirúrgicas livres) e melhora na qualidade de vida, atentando-se sempre para a ressocialização. Palavras-chave: Fibromatose Agressiva; Toracoplastia; Desarticulação; Reabilitação; Socialização.

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INTRODUCTION

The desmoid tumor (DT), also known as aggressive fibromatosis, represents 0.03% of all neoplasms and 3% of soft tissue tumors. Is that neoplasia that stretches

into musculoaponeurotic tissues, penetrates joint capsules, infiltrates along fascial planes, and invades adjacent neurovascular structures.¹ It is characterized by low metastatic potential and exuberant locoregional growth and high rate of local recurrence after surgical resection.

This is the report of a previously healthy young man, with a desmoid tumor in the hemithorax right scapular region, with the treatment proposal of thoracic interscapular disarticulation and right hemithorax ectomia aiming at R0 resection (with free surgical margins), with subsequent evaluation by the clinical oncology team for the initiation of adjuvant treatment.

CASE DESCRIPTION

This 23-year-old patient, male, leucodermic, previously healthy, carrier of a voluminous mass in the anterior thoracic wall, with the evolution of approximately one year. He reported having postponed medical appointments for fear and fear of being subjected to extensive surgery that would compromise his right upper limb. He was asymptomatic, which collaborated to seeking a late treatment, which occurred when the mass had great proportions, aesthetically damaging.

Propaedeutic was started in the Basic Health Unit and subsequently being referred to the Thoracic Surgery Service in the Mario Penna Institute /Luxemburgo Hospital.

He presented a voluminous mass in the right hemithorax (Figure 1), with biopsy on 12/13/11 showing fusocellular low-grade sarcoma. The computed tomography of the thorax showed bulky solid heterogeneous and expansive process of the anterior wall and right thoracic cavity, measuring 25 x 22 x 15 cm, associated with the deformity and erosion of the right ribs, partial ipsilateral lung atelectasis, compression and contralateral mediastinal deviation; little expansion of the left lung; trachea and previous main bronchi with normal gauge. Heart with normal dimensions.

Tumor resection was initially proposed with the attempt to preserve the limb. The patient was advised about the possible interscapular disarticulation and hemithorax ectomia and agreed with the proposal, being forwarded to the Psychology Service.

On 1/5/12 was submitted to exploratory thoracoscopy, which did not reveal pleural implants and the mediastinum was tumor free.



Figure 1 - Pre-operative aspect of the lesion.



Figure 2 - Reconstruction of the thoracic wall with marlex screen and methyl methacrylate.



Figure 3 - Reconstruction of the thoracic wall with marlex screen and methyl methacrylate.

On 1/17/12, during the perioperative period, invasion of mediastinum and infiltration of the subclavian artery and vein and brachial plexus by the tumor were observed. Toracectomy, resection of tumor in the mediastinum, amputation of the right arm, scapulectomy, and reconstruction of thorax wall with marlex screen and methyl methacrylate and preparation of fasciocutaneous retail were performed (Figures 2 and 3).

The patient was transferred to the intensive care unit, hemodynamically unstable, requiring the use of amines, being discharged to the infirmary on 1/20/12.

Good postoperative evolution with discharge on 1/30/12 and transfer to the Oncology clinic.

In the late postoperative period, he evolved with infection of surgical wound requiring removal of the methyl methacrylate prosthesis.

In a follow-up, the anatomopathological analysis of the surgical piece exhibited compromised margins – medial margin next to thoracic spine and new chest computed tomography showed tumor remnant along the second posterior costal arc - next to the spine.

The immunohistochemical study accused negative desmin-D33, negative smooth muscle actin 1A4, negative S-100 polyclonal protein, negative CD34-QBEnd10 clone, positive beta catenin 14, the results of which characterize “desmoid type fibromatosis.”

The use of tamoxifen was initiated because the standard chemotherapy scheme does not provide good response to these types of tumors.

The patient is in good physical and psychological recovery. He began rehabilitation for his re-integration into the labor market (Figure 4) and is in psychological, physical therapy, and surgical oncology monitoring.



Figure 4 - Post-operative aspect.

DISCUSSION

Desmoid tumors, in most described cases, are associated to familial adenomatous polyposis (PAF), especially in the clinical variant called Gardner syn-

drome, in which, besides the polipsecolonico, cutaneous tumors occur concomitantly (lipomas, epidermoid cysts), osteomas, dental malformations, and congenital hyper pigmented retinopathy.

It is subdivided according to their anatomical location in intra-abdominal, abdominal, and extra-abdominal. In its thoracic presentation, it can affect only the wall in 47% of the cases or invade adjacent structures in 53% of the cases.²

Its main manifestation is the presence of a palpable mass, thoracic pain and, depending on the invasion of adjacent structures, involvement of the amplitude of movements of the ipsilateral limb.^{2,3}

The treatment consists of surgical resection with expanded margins since its rapid growth can determine invasion of adjacent structures, even if in the case of tumor of benign behavior.³

The radio and chemotherapy and the use of non-cytotoxic drugs (AINEs, hormonal therapies, and biological agents) must also be considered, especially in cases of unresectable tumors.³

In the era of minimally invasive surgeries, it seems controversial to advocate major surgeries perceived as aggressive and mutilating.

Young and previously hidig patients are able to tolerate large interventions, with good recovery. Another point to note is a big breakthrough in the area of rehabilitation. This allows for patient's rehabilitation in the social and professional environment.

The multi-professional team and the synchronized and cooperative actions among the areas involved provide quality of life for the patient, often better than that before the surgery. In some cases, the tumor is more socially exclusionary than the physical disability caused by the amputation of a body segment.

The society has evolved to accept physical disabilities and mutilations. New apparatuses and public and social works reintegrate physical handicapped into everyday life. However, cancer is still stigmatized. Patients with extensive lesions are discriminated against and left on the sidelines because of the physical aspect.

In this case, the patient presented extensive chest lesion, which in addition to extremely painful and debilitating, excluded him from social conviviality and routine activities.

The complete resection of the lesion was chosen; however, the medial margin next to the thoracic spine was microscopically compromised. The surgery aimed at reducing the risk of death because the tumor was already starting to have a mass effect on the mediastinum.

In fact, the resection was not curative, which was once considered R1 - compromised microscopic margins. However, it succeeded in the proposal of improvement in quality of life, providing active and integrated life as the essence of the treatment.

CONCLUSION

The desmoid tumor treatment still constitutes a challenge. These tumors are relatively rare, composed of heterogeneous histological types and presenting diversified biological behaviors.

Despite these data, the interscapular disarticulation and hemitorax ectomia showed to be the only potentially curative therapeutic option.

The presented case, even after a mutilating and radical operation, the R1 resection was still consid-

ered, with the requirement of adjuvant therapies for the control of the disease. Considering that this was a young patient, higid, with a slow-growing tumor, despite its size, this therapeutic tactic seems the most effective regardless of its impact to the patient's life.

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