Case report

Intrathoracic desmoid tumor with invasion of the great vessels

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Abstract

A desmoid tumor of the mediastinum was diagnosed and treated in a 35-year-old white male who presented with a right supraclavicular mass. He was treated with resection, which involved several vascular structures, requiring multiple vascular reconstructions followed by post-operative radiotherapy. The authors concluded that, when located in the mediastinum, the invasive character of such tumors and its tendency to recur may pose a considerable surgical challenge, requiring careful pre-operative planning and long term post-operative follow-up. The role of radiation therapy is limited to the control of local recurrences.

Keywords: Desmoid; Fibromatosis; Mediastinum; Surgery

1. Introduction

Desmoid tumors, known as fibromatosis, are locally invasive tumors included in the spectrum of the low grade soft tissue sarcomas. These neoplasms account for less than 0.03% of all tumors [1], usually arising in the musculofascial of the abdominal wall. Desmoids of the chest wall and shoulder girdle occur in 40% of the instances. The radicality of the resection is paramount for the successful outcome since desmoid tumors tend to recur locally. We report a case in which resection required complex vascular reconstructions.

2. Case report

A 35-year-old white male was referred to our division because of a right chest pain starting 6 weeks before his visit. He also noticed a right supraclavicular mass and neck vein dilatation within 2 weeks prior to his hospital admission. Upon physical examination, there was a hard right supraclavicular mass adherent to the deep muscular structures of the neck. There was a fixed vein dilatation on both sides of the neck. Chest roentgenograms showed an anterior-superior mediastinal mass. Computed tomography (CT) scan showed a solid cervical-mediastinal infiltrative tumor extending from the posteromedial aspect of the right thoracic inlet to the lateral wall of the aortic arch and superior vena cava (SVC). Magnetic resonance imaging (MRI) of the chest (Fig. 1) and digital angiography demonstrated a circumferential involvement of the brachiocephalic artery extending to the right common carotid and subclavian artery without signs of intraluminal invasion. A cavogram showed obstruction of the left innominate vein, right subclavian, internal jugular veins and SVC which was obstructed proximally. Two percutaneous fine needle aspirations were not diagnostic. Open biopsy of the supraclavicular mass revealed a desmoid tumor (fibromatosis). A fiberoptic bronchoscopy was normal. Upon completion of the pre-operative work-up, the patient was taken to the operating room and a right oblique cervical incision was combined with a median sternotomy providing an excellent exposure of the tumor. It originated in the medial aspect of the muscular fascia of the deep cervical muscles within the thoracic inlet, invading the outer aspect of the vascular structures in addition to the right phrenic and vagus nerves. En-bloc dissection of the tumor was carried out including its attachments to the deep cervical and paraspinal muscles, vascular structures, right vagus and phrenic nerves close to the pericardium. The vascular reconstructions were then carried out...
In order to minimize the time of cerebral ischemia, we elected to start bypassing the brachiocephalic artery with a 10 mm Dacron woven straight graft extending from the origin of the brachiocephalic artery to the common carotid, followed by an 8 mm Dacron woven interposed between the distal cut end of the right subclavian artery and anastomosed end-to-side to the other graft. The tumor was then removed en-bloc with the superior portion of the SVC, left innominate vein, right phrenic and vagus nerves and the stellate ganglion, leaving free macroscopic surgical margins from all aspects of the tumor mass. The right subclavian and internal jugular veins were ligated. An 18 mm compressed bovine pericardial graft was anastomosed end-to-end between the left innominate vein and the SVC (Fig. 2). Chest tubes were placed and the incisions were closed. Early post-operative outcome was uneventful and the patient was discharged on the 12th post-operative day with a mild parestesia in the right arm due to manipulation of the brachial plexus (C8–T1 nerve roots) and permanent Horner’s syndrome on the right side and right diaphragmatic paralysis. He was re-admitted 2 weeks later due to an acute thrombophlebitis of the right arm, was treated with penicillin and systemic anticoagulation being discharged 9 days later on coumadin. The pathology specimen confirmed the primary diagnosis of desmoid tumor and showed microscopic invasion of the surgical margin in the medial aspect of the thoracic inlet adjacent to T3–T4 thoracic vertebrae. The patient was then referred to adjuvant radiotherapy (50 Gy) in the mediastinum. Coumadin was discontinued 6 months later. Despite some degree of ventilatory restriction due to the right phrenic paralysis and hoarseness secondary to the resection of the vagus nerve the patient has been otherwise asymptomatic and has resumed his activities as a full time office worker. Chest CT scans and digital angiography performed 1 year after resection showed patency of all grafts. Four years after surgery an occlusion of the SVC graft was detected on a follow-up CT scan, however the patient was asymptomatic and has remained disease free 6 years after treatment.

3. Discussion

Expected 5-year survival for resected chest wall desmoid tumors is 93% despite a high local recurrence rate (29%) [2]. In our patient the tumor originating in the inner aspect of the chest wall and mediastinum invaded vascular and neural structures. Since desmoid tumors usually do not metastasize, the treatment focuses on local control only achieved by wide resection with tumor-free surgical margins. Therefore, one has to plan the surgical approach carefully in order to accomplish a radical resection which may often require parietal and vascular reconstructions. We elected to start resecting the tumor invading the innominate artery and to carry out its reconstruction first in order to shorten the time of cerebral ischemia as much as possible. Thereafter resection and reconstruction of the right subclava-
vian artery, followed by the left innominate vein and SVC were performed. The arterial grafts remained patent, however the cause of the late occlusion of the SVC graft remains unclear. We hypothesize that the early thrombophlebitis might have played a role in this regard. Other treatment modalities for desmoid tumors include radiation and chemotherapy. Patel and Evans [3] reported their experience with a doxorubicin-dacarbazine regimen and found a complete/partial response in 50% of the patients treated. Following the post-operative pathology findings of microscopic positive resection margin in the mediastinal aspect of the tumor, we elected to send the patient to radiotherapy since the extent of the resection was already considered maximal. Sherman and Romsdahl [4] used this approach and achieved local control of the disease in 16 out of 22 patients with positive surgical margins with doses varying from 50 to 76.2 Gy. Dosios et al. [5] recommended adjuvant radiotherapy for control of local recurrences. We conclude that the successful outcome is based on careful pre-operative planning and long term post-operative follow-up, given the recurrent pattern of such tumors. Adjuvant radiotherapy may play a role for local control when positive surgical margins are present as well as for the treatment of recurrences for selected patients whenever another resection is not deemed feasible.

References