Case Report

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The Impact of Radiotherapy in the Treatment of Recurrent Desmoid Tumor of Neck: A Case Report with Literature Review

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Abstract

Background: Extra-abdominal desmoid tumor of the head and neck in particular commonly infiltrate vital neurovascular structures. we report a case of a large and recurrent extra-abdominal desmoid tumor causing significant morbidity and who was treated successfully by radiotherapy.

Case presentation: A 34-year-old male patient presented with a history of right neck mass for 15 months, which was gradually increasing in size. Cervical CT scan revealed a large mass of the posterior soft tissues of the neck extended to the soft tissues of the posterior thoracic wall. The CT density of the mass was that of fat tissue. It was measuring 12.4 × 85.9 mm and enhanced after injection of the contrast product. A biopsy was taken. The histopathological findings were suggestive of desmoid tumor. This case was discussed in multidisciplinary oncological team and radiotherapy was planned. The surgery was proposed but not performed due to the recurring character of the tumor and the risk of a high postoperative morbidity. After 4 weeks of treatment, the clinical evaluation showed a good response.

Conclusion: Alternative treatment modalities such as primary radiotherapy may be preferable to mutilating surgery.

Keywords: Desmoid tumor; Neck; Surgery; Radiotherapy

Introduction

Desmoid tumor (DT) or deep-type musculoaponeurotic fibromatoses is a rare infiltrative fibroblastic tumor that present complex management considerations [1]. Extra-abdominal desmoid tumor of the head and neck in particular commonly infiltrate vital neurovascular structures including the brachial plexus emphasizing the importance of function-sparing surgery. In general, DT have a female predominance while the highest incidence arises among patients between the ages of 15 to 60 years old [2]. In light of these trends, we report a case of a large and recurrent extra-abdominal desmoid tumor causing significant morbidity and who was treated successfully by radiotherapy.

Case Presentation

A 34-year-old male patient presented with a history of right neck mass for 15 months, which was gradually increasing in size. 3 years later, he reported the same symptoms of growing cervical mass treated by surgical resection (we don't have surgical report or any data). On examination, there was a neck swelling involving the left postero-lateral aspect of the upper neck shown in **Figure 1**.

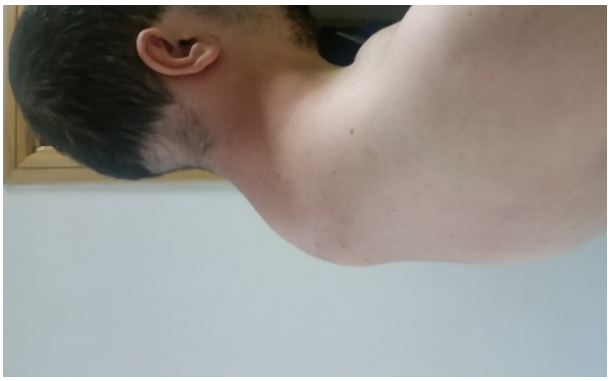

Figure 1 Lateral view of the patient showing the desmoid tumor of neck.

which was hard and fixed to the skin and deep neck structures. Cervical CT scan revealed a large mass of the posterior soft tissues of the neck extended to the soft tissues of the posterior thoracic wall. The CT density of the mass was that of fat tissue. It was measuring 12.4 × 85.9 mm and enhanced after injection of the contrast product. It has bumpy contours and blurred edges shown in **Figure 2.**

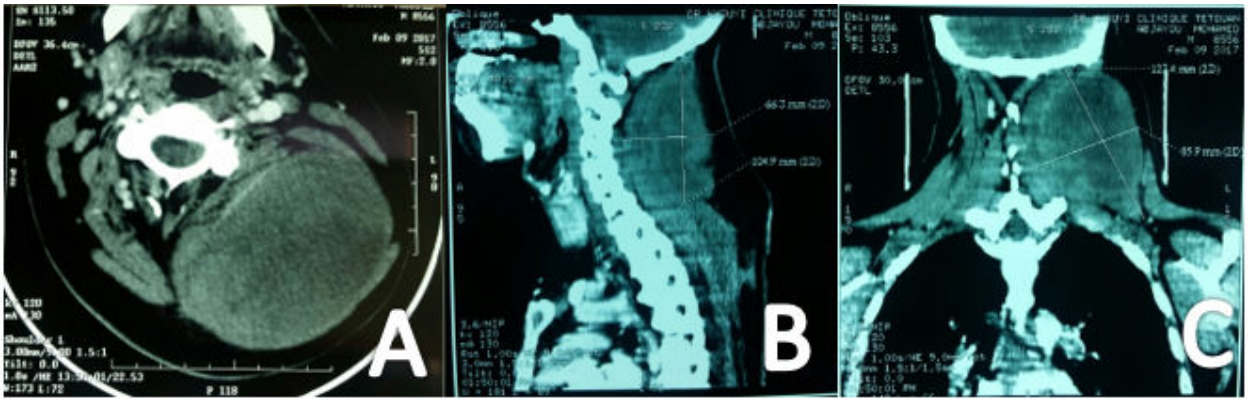

Figure 2 Different planes of CT scan showing the cervical mass (A: Axial plane, B: Sagittal plane, C: Frontal plane).

This mass invaded the scalene muscles with predominance on the right. Below it extends to the paravertebral muscles. There is a tumor vascularization from the arterial branches of the brachio-cephalic trunk. The carotid arteries were free shown in **Figure 3**.

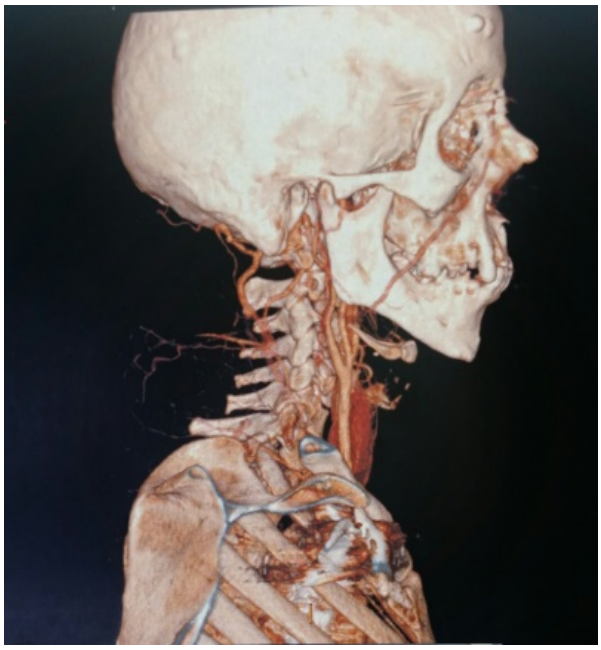

Figure 3 Image of Angio CT scan showing tumor vascularization from the arterial of the brachio-cephalic trunk. The carotid arteries were free.

A biopsy was taken. The histopathological findings were proliferating stellate to spindle cells arranged in long fascicles or whorling patterns with bland nuclear features and dense keloid-like collagen in areas. The cells did not show nuclear atypia or hyperchromasia shown in **Figure 4.**

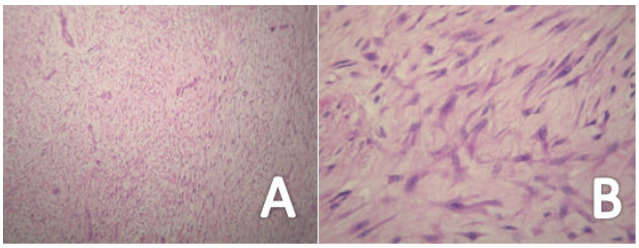

Figure 4 Histopathologic examination of the biopsy under low (A, 4x) and high (B, 10x) power magnification reveals an elongated fascicle with uniform fibroblastic spindle cells, mild to moderate cellularity without atypia or pleomorphism, consistent with a benign desmoid fibroma (Hematoxylin and Eosin stain).

Immunohistochemistry stains from pan cytokeratin, EMA, PS100, CD34 were negative while KI67 and beta-catenin stains were positive shown in **Figure 5.**

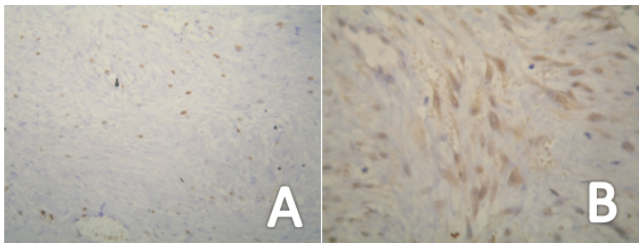

Figure 5 Immunohistochemistry stains: KI67 (A, x2) and beta-catenin (B, x4) positivity.

The diagnosis of desmoid tumor of neck was retained. This case was discussed in multidisciplinary oncological team and radiotherapy was planned. The surgery was proposed but not performed due to the recurring character of the tumor and the risk of a high postoperative morbidity. External beam radiotherapy at a radiation dose of 2 Grays (Gy)/fraction given five times weekly (Monday to Friday) to a dose of 50 Gy. Overall treatment time was 5 weeks. Patient completed the

treatment without any major untoward event. After 4 weeks of treatment, the clinical evaluation showed a good response shown in **Figure 6**. A radiologic evaluation by cervical CT scan is planned after 3 months.

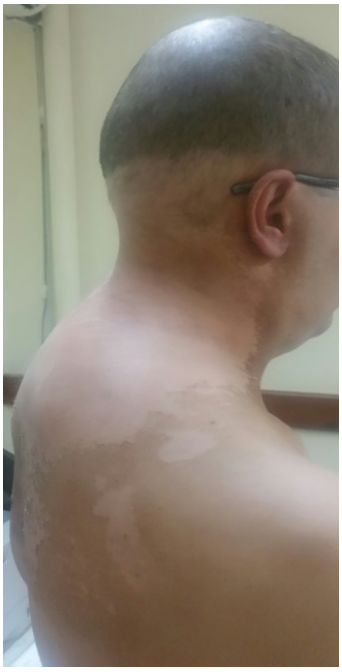

Figure 6 Lateral view of patient after 4 weeks.

Discussion

DT is characterized by local aggressiveness with an approximate 20 % local recurrence rate, but without metastasis [3]. The annual incidence of DF is presumed to be 0.2 to 0.4 per 100,000 individuals [4]. Among cases of DT, 7 to 15 % of tumors occur in the head and neck [5]. Wide surgical resection with clear margins has long been advocated as the first-line treatment of choice for desmoid tumor [6,7]. But there are situations in which alternative methods or adjuvant therapy should be considered. For example, debilitated patients or cases when the patient refuses radical resection. Because of the difficulty in determining the actual margins of these extensive lesions at surgery, positive margins are not unusual. In this situation, in recurrent or inoperable disease, radiation therapy should be considered [7]. The total dose applied should be above 50 Gy [5]. Several reports have demonstrated that radiotherapy can achieve complete and long-term regression of the disease [7]. Although it would be difficult to realize because of the rarity of these tumors, the contribution of radiotherapy to the treatment of desmoid tumours can only be answered by a prospective randomized clinical trial in a defined patient group [5]. The prognosis depends upon the initial treatment. If this is inadequate, the tumor will recur one or more times and will often progress [7].

Conclusion

Alternative treatment modalities such as primary radiotherapy may be preferable to mutilating surgery. Considering all the afore mentioned, it seems obvious that DT of the neck present a therapeutic challenge and require an individualized approach.

Conflict of Interest

The authors declare that they have no conflict of interest that competes with any of the contents of the manuscript.

Ethics Approval and Consent to Participate

Written informed consent was obtained from the patient and her family for publication of this case report and any accompanying images.

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