Case Report

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Journal of Clinical Radiology and Case Reports

2020
Vol. 4 No. 3

Cervical Desmoid Tumor: About a Case and Review of Literature

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Received: November 09, 2020; **Accepted date:** November 23, 2020; **Published date:** November 30, 2020

Citation: Agbanglanon DPE (2020) Cervical Desmoid Tumor: About a Case and Review of Literature. J Clin Radiol Case Rep Vol.4 No.3

Abstract

Desmoid tumors (DTs), also called well-differentiated fibromatosis, are circumscribed but not histologically encapsulated benign neoplasms infiltrating muscles and fascia. They tend to be locally infiltrating and recurrent but without metastatic potential. These tumors are most often located in the abdominal and thoracic wall, cervical localization is rare.

We report on a 32-year-old patient who presented with a laterocervical desmoid tumor (DT) that was recused from surgery and who received neo-adjuvant chemotherapy with lesional stability and adjuvant radiotherapy.

Keywords

Desmoid; Fibrotic tumor; Neck; Surgery; Radiotherapy.

Introduction

Desmoid tumors (DTs), also known as aggressive fibromatosis or well-differentiated fibromatosis, are benign soft tissue tumors originating in deep musculoaponeurotic structures. They are infiltrating and recurrent fibrous proliferations, but without metastatic potential. They are rare tumors as they account for less than 0.03% of all neoplasias [1]. Exceptional cervical localizations are particularly aggressive [2]. Computed tomography (CT scan) and especially magnetic resonance imaging (MRI) are the tests of choice to clarify a possible locoregional extension. We report the case of a patient presenting a DT of the right laterocervical region that was recused from surgery.

Case Review

A 32-year-old, female, with no significant past medical history, admitted to our institution with a relatively slow growing tumor which started 14 months earlier, painless right laterocervical swelling, evolving in a context of apyrexia, preservation of the general state and without other associated signs. On physical examination, a large, fixed right laterocervical swelling about 7 cm wide was found, of firm consistency, painless, adherent to the deep plane and with no inflammatory signs in sight **(Figure 1)**. The rest of the examination showed no particular sign, there was no neurological deficit. A cervico-thoraco-abdominal CT scan was performed and was in favor of a laterocervical mass **(Figure 2)**. A biopsy showed a spindle cell tumor, which was primarily suggestive of a DT. On immunohistochemistry, the tumor cells expressed focal actin smooth muscle (ASM), negative for antibodies to PS100 and epithelial membrane antigen (EMA) **(Figures 5,6)**. MRI showed a poorly limited laterocervical tissue lesion process of approximately 70 x 60 x 112 mm with irregular contours in iso-signal T1, discrete hypersignal in heterogeneous T2 strongly enhanced after contrast injection. This tumor infiltrated the scalene muscles, invaded the long muscles of the neck, the splenium of the neck, and came into contact with the right lateral chest wall with no sign of invasion of bone or lung opposite **(Figures 3 and 4)**. The case was presented at a multidisciplinary consensus meeting (MCM) and the decision was that the tumor was not surgically resecable. The patient received 9 courses of neoadjuvant Navelbine-based chemotherapy. An evaluative MRI scan performed showed lesional stability. Adjuvant radiotherapy was decided. Treatment with photons was performed as intensity modulated radiotherapy, IMRT in VMAT technique at the discretion of the according to radiation oncologist. Gross Tumor Volume (GTV) included the gross tumor based on CT and MRI imaging. The Clinical Target Volume (CTV) was defined as GTV plus surrounding areas at risk for containing microscopic disease. The CTV included the GTV aiming at a margin of 2 cm, depending on the location and anatomy. The CTV margins were smaller if the GTV was adjacent to the organ at risk. The PTV margin was 5 mm in all directions. The dose of radiotherapy was 56 Gy/fraction in 28 fractions at 2 Gy per fraction to the isocentre, using 6 MV photon and CT scan planning. It should be noted that the right brachial plexus was in full tumor volume. The dosimetric constraints of the organs at risk were respected with a tumor volume was 214.84 cm3. Treatment should be once a day, 5 days a week in five and a half weeks.

The early evolution after radiotherapy was marked by a stable lesion. There was no neurological disorder in the homolateral thoracic limb.

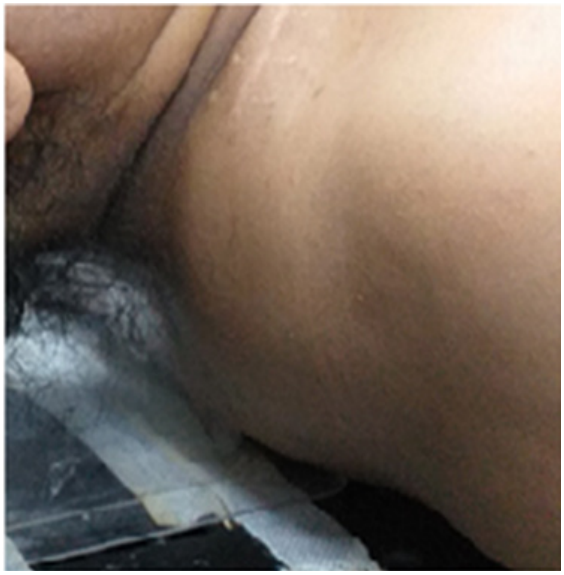

Figure 1 Right cervical tumor

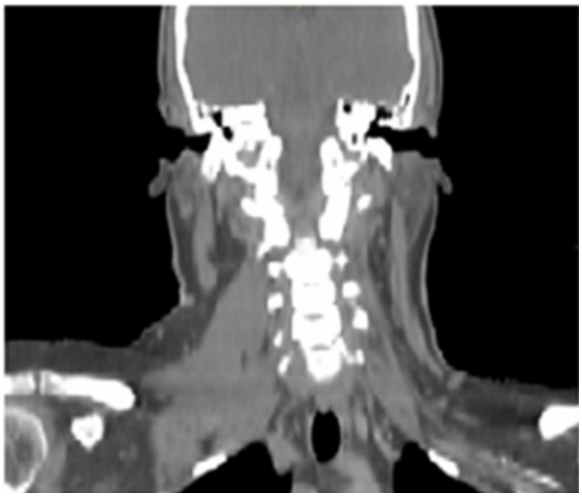

Figure 2 Frontal CT scan of the right cervical desmoid tumor

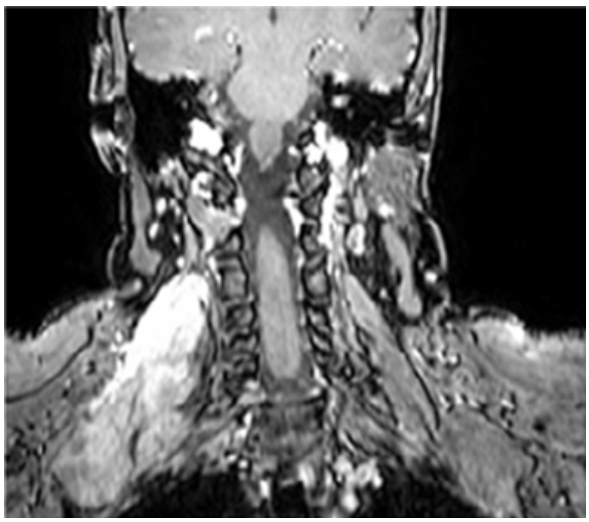

Figure 3 Frontal MRI sequence T1 with injection of the right cervical desmoid tumor

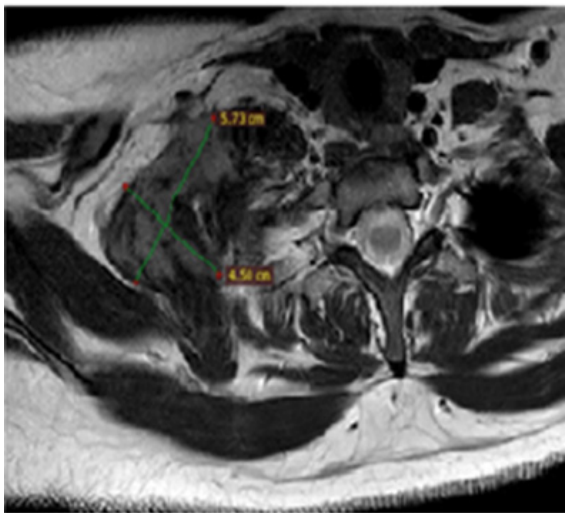

Figure 4 Coronal MRI sequence T2 of the right cervical desimoid tumor

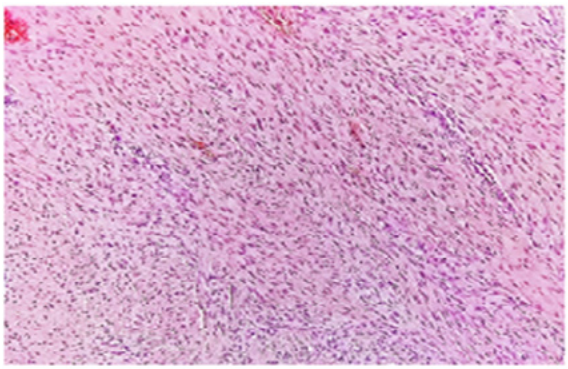

Figure 5 Proliferation of spindle-shaped cells corresponding to fibroblasts and myofibroblasts, arranged in long divergent bundles,HEx4

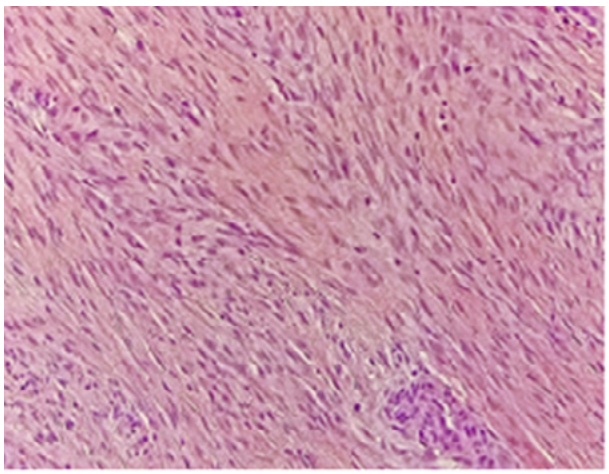

Figure 6 Cells do not show cytonuclear atypia or mitosis, HEx20

Discussion

The desmoid tumor (DT) also called aggressive fibromatosis is a rare pathology [3], it is part of the deep fibromatosis integrated in the group of soft tissue tumors. It accounts for less than 0.03% of all neoplasias, with an incidence of 2 to 4 new cases per 100,000

per year [4,5]. They can occur at any age [6,7] in both genders, but especially in young women between 20 and 40 years of age [4]. While most cases are sporadic, 2% are of genetic origin, by mutation of the APC gene, and are part of Gardner syndrome associated with familial adenomatous polyposis (FAP) [4,8]. They are generally unique, but multifocal localizations have been described [6]. Cervical localizations are particularly aggressive: they can lead to destruction of the adjacent bone, erosion, and even compression of the vascular nerve bundle and of the laryngotracheo-esophageal visceral axis [2], which has not been observed in this patient. Several hypotheses have been made to explain the occurrence of these tumors, namely traumatic origin, hormonal influence, and genetic and hereditary factors [6].

They appear in the form of a poorly limited mass of slow growth and are generally not very painful. They occur mainly in the waist and neck regions [7]. They adhere to the deep, never to the superficial plane, are firm in consistency, and are painless in the majority of cases [9], as in this patient's case. Depending on the situation, they may cause discomfort to mobility or signs of neurological compression [7], which is not the case in our patient. This typical clinical aspect may evoke the diagnosis before further examination. Radiologically, it is a soft tissue mass that often erodes adjacent bone tissue. CT and especially MRI are the tests of choice. They make it possible to assess the limits of the tumor and its relationship with the vasculonervous bundle. On T1-weighted images, DTs are hypo or iso-intense to the muscles, while on T2-weighted images they are hyper-intense. With gadolinium contrast, the DT shows moderate accentuation with hypo-intense bands reflecting collagen bundles. The diagnosis of certainty is based on anatomopathology after surgical biopsy. Histological examination confirms the diagnosis and seeks to rule out a low-grade fibrosarcoma, a reactive fibroblastic proliferation or a nodular fasciitis. The differential diagnosis includes rhabdomyosarcoma, malignant histiocytoma, neurofibroma and mesenchymal tumors [3]. Histological evidence by biopsy is considered essential by some teams [10]. However, this attitude is controversial¬. Indeed, the traumatic nature of a surgical biopsy could accelerate tumor growth, with a risk of complications. However, when the tumor cannot be removed, as in our clinical case, the biopsy helps to make the diagnosis in order to better consider the therapeutic management [7].

Despite the inadequacy and difficulties of surgical treatment, wide excision remains the first-line treatment recommended by the majority of authors [11,12]. However, it is not always possible because of the size of the tumor, the invasion of the vasculonervous bundle and the poorly limited aspect of the tumor, as is our case. The recurrence rate after surgery is on average over 50% [3,13]. It can reach 90%, especially for children [11,13]. For some authors [3,8,13], this rate is higher in the case of incomplete removal, while for others [12], the quality of the initial removal has no influence. In view of the high risk of recurrence, empirical second-line treatments have been recommended, including radiotherapy, hormone therapy and chemotherapy [3]. Radiotherapy provides a definite benefit in cases of residual disease or in inoperable sites [14]. In order to reduce the recurrence rate and the functional risks of surgery, some authors propose the use of radiotherapy in a systematic way [15]. The response to radiotherapy in these cases is very slow but presents a good local control rate, which can be as high as 81.5%. However, neurological and therefore functional complications are not uncommon, although they are rarely observed for doses of less than 60 grays [16]. The role of radiotherapy in the prevention of recurrences in the event of positive recurrence is controversial [6,8]. In young subjects, its indication should be discussed taking into account the risk of radiation-induced tumors [3]. The combination of radiotherapy and hormone therapy is a good alternative that provides objective answers in inoperable DT [6]. The slow evolution of these tumors does not suggest a high chemosensitivity; however, it has been tested in inoperable relapse situations with some significant results [9].

Evolution is always unpredictable. Spontaneous regressions have been reported without any treatment or after partial resection [14], as well as tumor stabilization. Although never metastasizing, DTs are characterized by a very high potential for recurrence, with the possibility of sarcomatous transformation [14].

Conclusion

DTs are an extremely rare entity whose diagnosis is complex. They often affect young subjects. Their evolution is slow and unpredictable. However, there is no clear consensus on their therapeutic management.

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