ONCOLOGÍA

ISSN: 2661-6653

DOI: https://doi.org/10.33821/732
Article / Artículo

Calcifying fibrous pseudotumor of the neck in a teenager female patient: case report

Pseudotumor fibroso calcificante de cuello en una paciente adolescente: reporte de caso

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Received: 08/01/2024
Accepted: 11/03/2024
Published: 30/04/2024

ABSTRACT
Introduction: Calcifying fibrous pseudotumor is a benign soft tissue tumor, appearing mainly in children and young adults between 20 and 30 years of age, still without a clear and defined etiology. It has a variable body distribution, being relatively uncommon in the neck. **Case report:** We present the case of a 17-year-old teenager female patient with a fast-growing neck tumor; its manifestation was moderate localized pain, excessive snoring, and progressive respiratory distress. **Treatment:** Complete resection of the tumor located in the hypopharynx was performed, which confirmed the histopathological and immunohistochemical diagnosis of calcifying fibrous pseudotumor. **Conclusion:** Calcifying fibrous pseudotumor of the neck is a benign pathology, rare, with non-specific symptoms, and most probably by the tumor compressing the surrounding tissues. Surgical resolution remains the gold standard for treatment. The prognosis after resection is good in the long term, with low recurrence rates. The diagnostic and therapeutic approach in this teenage patient is discussed compared to that described in the literature.

Keywords: Calcifying fibrous pseudotumor, psammomatoid calcifications, benign neck tumor, cervical tumor, teenager.

RESUMEN
Introducción: El pseudotumor fibroso calcificante es un tumor benigno de tejidos blandos que aparece principalmente en niños y adultos jóvenes entre 20 y 30 años de edad, aún sin una etiología clara y definida. De distribución corporal variable, siendo relativamente poco común en el cuello. **Caso clínico:** Se presenta el caso de una paciente adolescente de 17 años con tumor en el cuello de rápido crecimiento que se manifestó con dolor moderado localizado, ronquido excesivo y dificultad respiratoria progresiva. **Tratamiento:** Se realiza resección completa del tumor localizado en hipofaringe, la cual confirma el diagnóstico histopatológico e inmunohistoquímico de pseudotumor fibroso calcificante. **Conclusión:** El pseudotumor fibroso calcificante de cuello es una patología benigna rara, con síntomas inespecíficos y muy probablemente, inducida porque el tumor comprime los tejidos circundantes. La resolución quirúrgica sigue siendo el Gold estándar (GS) en cuanto al tratamiento. El pronóstico postresección es bueno a largo plazo, con tasas bajas de recurrencias. Se discute la aproximación diagnóstica y terapéutica en un paciente adolescente comparado con lo descrito en la literatura.

Palabras Clave: DeCS: Pseudotumor fibroso calcificante, calcificaciones psamomatoides, tumor benigno del cuello, tumor cervical, adolescente.

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How to cite: Bombón MF, Criollo Vargas E. Calcifying fibrous pseudotumor of the neck in a teenager female patient. Oncología (Ecuador). 2024;34(1): 44-51. https://doi.org/10.33821/732

1. Introduction

Calcifying fibrous pseudotumor is a rare benign soft tissue tumor with distinctive histologic features [1]. It is characterized histologically by abundant collagenized tissue, with a focal lymphoplasmacytic infiltrate and psammomatous and dystrophic calcifications [2]. It is more frequent in children and young adults [3]. They are found in numerous body sites, most often in the gastrointestinal tract or subcutaneous soft tissue; however, they are relatively uncommon in the neck [1]. The cause and pathologic mechanisms are unknown [3]. In most cases a single lesion is described, although there are cases of multiple lesions. Its diagnostic approach is clinical and by imaging. It is necessary to have a histopathological specimen to confirm the diagnosis [4]. The aim of this article is to report the case of a calcifying fibrous pseudotumor in the neck of a teenager patient.

2. Case Report

We present the case of a 17-year-old teenager female, with no significant pathologic history, who presented an apparent neck tumor causing moderate pain, excessive snoring for about 4 months and that was worse in recent weeks accompanied by progressive respiratory distress. No neurological or digestive symptoms or signs were evidenced. The patient initially underwent a CT scan of the face and neck, without a definitive diagnosis, which reported paratracheal and retrotracheal tumor with calcifications inside, displacing and partially obliterating the larynx and trachea with regular contours measuring 4 x 11 cm.

On physical examination, there was an increase in cervical volume occupying the entire central topography of the neck, soft to palpation. The oropharynx was palpable bulging, totally compromised by a hard tumor.

2.1. Diagnostic workshop

The patient was evaluated by the Head and Neck Surgery Department, under the presumptive diagnosis of tumor of uncertain behavior of the neck, some complementary examinations were requested, including paraclinical laboratory studies, nasofibrolaryngoscopy, CT of the face, neck, thorax, and ultrasound-guided cytopuncture of cervical tumor.

In the paraclinical laboratory studies, it was evidenced that the hemogram, blood biochemistry, coagulation times performed in the Clinical Laboratory Department of SOLCA-Guayaquil, all stayed in normal parameters.

The nasofibrolaryngoscopy showed tumor of nasopharynx limit with oropharynx, which occupies the entire oropharynx and reaches the hypopharynx, thus displacing the larynx towards the anterior. Due to the size of the mass, it is not possible to access the supraglottis.

The report of the CT of the face and neck with intravenous contrast identifies tumor lesion of defined contours, heterogeneous with soft tissue density and irregular calcifications inside, measuring 69 mm in its major axis in axial plane and 117 mm in its major axis in sagittal plane, with no enhancement after intravenous contrast. On its former margin it contacts and compresses the pharynx, larynx, esophagus and thyroid gland, predominantly in the left lobe. Its later margin involves the prevertebral space, without apparent infiltration of vertebral foramina or bony structures, its lateral margins contact and displace the carotid and jugular veins, without causing infiltration of them. Permeable nasopharynx, with preserved morphology, oropharynx occluded almost completely by the described tumor lesion, no cervical adenopathies are observed (Figure 1).

Chest CT reports: cervical prevertebral tumor lesion, extending to the thoracic operculum, displacing the thyroid anteriorly without infiltrating it and deforming the posterior wall of the trachea, compressing the esophagus.

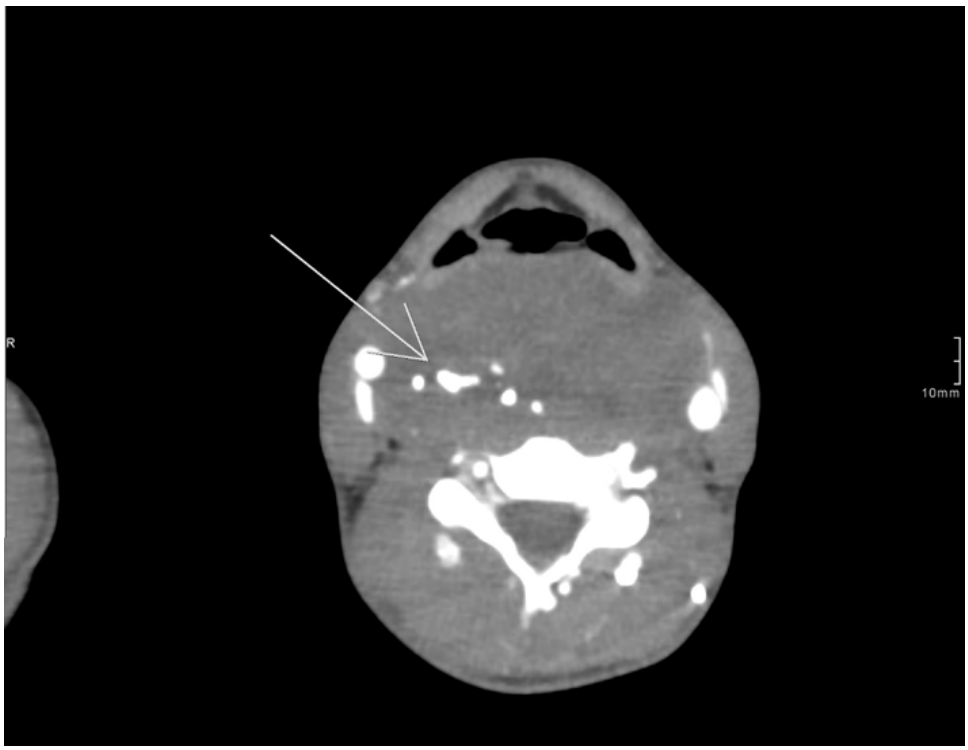

Figure 1. Axial CT scan of face and neck with intravenous contrast with evidence of tumor lesion of defined contours, heterogeneous with soft tissue density and calcifications inside, no enhancement after intravenous contrast.
Source: Department of Radiology and Imaging. SOLCA - Guayaquil

Ultrasound-guided cytopuncture of cervical tumor was neither assessable nor conclusive, since the sample consisted exclusively of erythrocytes with traces of hemolysis and scattered leukocytes. Subsequently, the patient was evaluated with the results of complementary examinations and when the patient did not have a conclusive pathological or cytological result and presented the initial symptoms with apparent exacerbation of the clinical picture (respiratory distress); it was decided to perform open tracheostomy + incisional biopsy of the oropharynx, as a prophylactic measure in case of a probable obstructive picture in the short or medium term.

The procedure is performed without apparent complications and the biopsy result reports paucicellular fibroblastic/myofibroblastic proliferation with spindle cells without significant atypia immersed in a dense collagenous matrix, accompanied by sporadic lymphocytes and scarce psammomatous calcifications. No areas of necrosis, anaplasia or elevated mitotic activity suggestive of malignancy are identified. The findings are suggestive of calcifying fibrous pseudotumor. Immunohistochemical findings rule out fibromatosis and KI:67 demonstrates a very low cell proliferation index.

In this context, the present case is evaluated and reviewed by the skin, sarcoma and soft tissue committee; subsequently by the pathology committee, who after a thorough and exhaustive analysis recommend increasing the immunohistochemical tests, which are detailed in Table 2.

The IHC tests confirmed the diagnosis of calcifying fibrous pseudotumor, with IgG4 positivity. Therefore, in view of the results, surgical resection was decided as the only treatment option. A face, neck and thorax MRI with contrast was requested to complement the imaging findings previously described and in order to define the morphological and anatomical characteristics of the lesion and its relationship with adjacent structures, as well as for the selection of the best surgical technique (Figure 2).

Table 2. Immunohistochemical findings. Source: Department of Pathology.

IHQ Test	Result
B CATENINA	Negative
SOX 10	Focal positivity in myofibroblasts
KI 67	1 %
ANTI-ALPHA SMOOTH MUSCLE ACTIN 1A4	Positive perivascular and in myofibroblasts
S 100	Negative
SYNAPTOPHYSIN	Negative
CHROMOGRANIN	Negative
CD-117	Negative
CD-34	Positive

Source: Departamento de Patología. SOLCA - Guayaquil

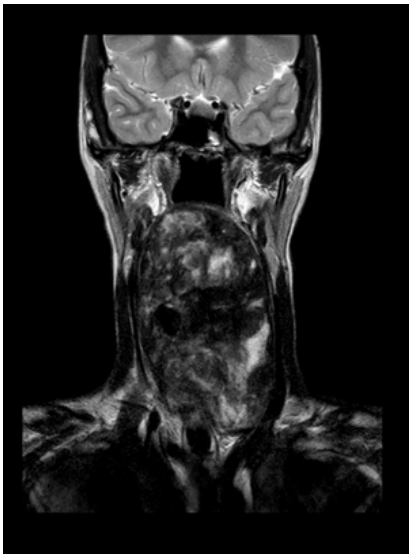

Figure 2. Simple and with contrast axial MRI of face and neck with evidence of extraluminal solid tumor mass in the oropharynx and hypopharynx space, which secondarily obliterates the lumen.
Source: Department of Radiology and Imaging. SOLCA - Guayaquil

The MRI of the face and neck reports: tumor lesion of solid aspect in the space of the oropharynx and hypopharynx, measuring 112 x 66 x 37 mm in its major axes, which secondarily obliterates its light in relation to its histopathological diagnosis of fibrous pseudotumor. Cervical adenopathies of probably inflammatory characteristics in levels I, II, and III there are adenopathies of preserved morphology, the largest measuring 8 mm. Inflammatory process in maxillary sinuses, sphenoidal and ethmoidal cells.

2.2. Treatment and evolution

The patient underwent an exploratory cervicotomy + left lateral dissection + tumor excision; the surgical procedure lasted 4 hours, with an approximate bleeding of 600 ml. A bilateral enlarged cervical incision was performed, finding intraoperatively a tumor in the hypopharynx region, with oval macroscopic characteristics, smooth surface, measuring approximately 8 x 10 cm (Figure 3). In addition, during the surgery, a left cervical lymph node level III, yellowish brown of elastic consistency, measuring 1.2 x 1 cm was isolated; left lateral dissection was performed. The vascular and nervous structures, such as parathyroid and recurrent laryngeal nerve were preserved.

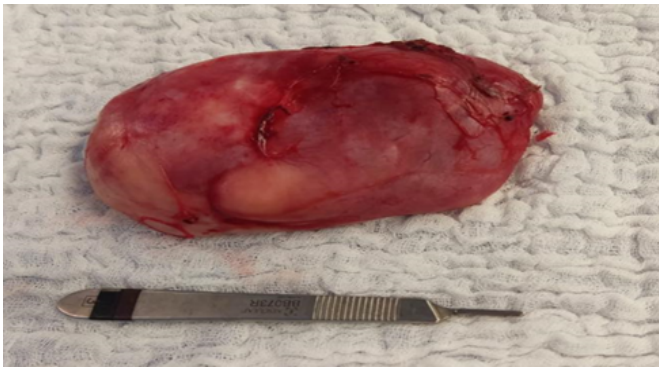

Figure 3. Intraperitoneal findings of hypopharyngeal tumor excision. Source: Solca-Guayaquil. Source: SOLCA - Guayaquil.

The pathology and immunohistochemical report of the surgical specimen indicated calcifying fibrous tumor.

The pathology study of the resected cervical lymph node revealed lymph node with sinusoidal histiocytosis, negative for malignancy.

Total excision of the calcifying fibrous pseudotumor of the hypopharynx was performed, without intraoperative complications, and there were no complications in the postoperative period. The patient was discharged 72 hours after surgery, with little bleeding due to the drains placed during surgery, which were removed after medical discharge, maintaining the tracheostomy, which was functional and in good condition. After the first week, the patient attended outpatient control in the Head and Neck area, where functioning tracheostomy, surgical wound in good healing process, no signs of infection, with relative improvement of the initial symptomatology were observed. Nasofibroscopy was performed and it reported good mobility of both cords. Respiratory exercises were indicated for eventual decannulation. The following week a successful decannulation was performed. Finally, an appointment was made for subsequent controls with results showing a sealed scar and no tumor recurrence. As this type of tumor is benign, the patient does not receive specific oncologic adjuvant treatment.

3. Discussion

The current WHO classification, since 2002, uses the term "calcifying fibrous tumor or pseudotumor" to describe a rare benign mesenchymal lesion [1,14]. Currently, the pathogenesis is uncertain. Possible etiologies include previous infection, trauma or surgical intervention [2].

This type of lesion is characterized by having a defined boundary, lacking a capsule and presenting a wide range of sizes. Microscopically, they present densely hyalinized hypocellular collagen with psammoma or dystrophic calcification and mononuclear inflammatory infiltrate [3,14]. It has been described mainly in children and young adults between 20 and 30 years of age, with little predisposition in women, even without a well-defined cause [4,5].

Calcifying fibrous pseudotumor has been documented in various anatomic locations, such as serosal surfaces, solid and tubular organs, and soft tissues [6]. The most frequently involved sites are the stomach (18%), small intestine (8.7%), pleura (9.9%), neck (6.2%), peritoneum (6.8%), mediastinum (5%), and mesentery (5%) [7]. In this context, there are very few neck cases reported in the literature, as most studies are sporadic case reports, with little or no symptomatology [1].

Patients usually have no specific symptoms, and when they have, these are atypical and most likely induced by the tumor compressing surrounding tissues. Tumors are often detected incidentally during imaging examinations or surgery [8].

Laboratory examination proved to be not so helpful in the diagnosis of calcifying fibrous pseudotumor [5]. The diagnosis is morphologic; therefore, immunohistochemistry could be of use in the differential diagnosis. The spindle cells are strongly and diffusely positive for Vimentin and Factor XIIIa and rarely positive for smooth muscle actin (SMA). Fibrous tumor immunoreactivity for CD34 has

been variably reported in the literature. An IgG:IgG4 ratio has been described in the plasma population, thus suggesting a potential association between IgG4-related disease [7,9].

The treatment of choice and best outcome for calcifying fibrous pseudotumor is surgical removal. Scientists agree that this type of tumor should be removed at the time of diagnosis. Few recurrences and no related deaths are reported [9]. However, other investigators suggest treatment with corticosteroids, but excision of radical tumors by surgery is the main method of treatment because postoperative histopathology and immunohistochemistry allow a definitive diagnosis to be obtained [10,12]. There are 2 types of excision recommended in the literature: open surgical excision and endoscopic excision. Open surgical resection or excision was performed in most cases; laparoscopic or minimally invasive surgery was also used in some cases [9,11]. CT and MRI provide a three-dimensional reconstruction of the lesion and define the morphologic features and their relationship to adjacent structures, which is important for surgical planning [1]. However, while imaging can play an important role in the diagnosis of many lesions, in this type of tumor particularly, the final diagnosis depends on the microscopic examination of the tissue performed in the histopathology study [1,5].

Cases of malignant transformation have not been reported in the literature [13]. Globally, studies have identified that in most cases no recurrences or metastases were observed, except in a few patients, including pediatric patients under 3 years of age, where there was a small recurrence within their follow-up period [15]. The prognosis is usually good with minimal associated morbidity and no reported mortality [13,15].

It is important to determine that the final and definitive diagnosis of calcifying fibrous pseudotumor is made among several similar entities, such as fibromatosis, synovial sarcoma, desmoplastic fibroblastoma, tendon sheath fibroma; exclusively with histological and immunohistochemical studies. Other studies dedicated to the identification of the exact pathogenesis of the tumor and evaluation of the age distribution of occurrence should also be performed to avoid misdiagnosis and unnecessary treatment [15].

4. Conclusions

Calcifying fibrous pseudotumor is a rare pathology, very difficult to diagnose, especially in teenager patients, such as the one in our clinical case. Symptomatology is nonspecific and the location variable, surgical management remains the cornerstone of treatment, with low recurrence rates and has a good long-term prognosis.

This particular case highlights the importance of considering calcifying fibrous pseudotumor as an entity to be taken into account when demonstrating cervical tumors under study, especially in teenager patients with non-specific symptoms. At present, since there are few cases described in the literature, appropriate diagnostic and therapeutic approach will allow proposing, evaluating, and following up future clinical cases as multicenter studies because these are huge opportunities for future medical research

5. Abreviaturas

IHC: Immunohistochemistry.

CT: Computed Axial Tomography

MRI: Magnetic Resonance Imaging

6. Administrative Information

6.1. Additional files

None declared by the authors.

6.2. Acknowledgments

We thank the patient and her family, who agreed to the dissemination of this scientific work.

6.3. Authors' contributions

Marco Fabricio Bombón Caizaluisa: Conceptualization, data curation, formal analysis, acquisition of funds, research, writing - original draft. **Emilio José Criollo Vargas:** Conceptualization, data curation, formal analysis. All authors read and approved the final version of the manuscript.

6.4. Funding

The investigators funded the study. The authors received no monetary recognition for this research work.

6.5. Availability of data and materials

Data are available upon request from the corresponding author. No other materials are reported.

6.6. Statements

6.6.1. Informed consent

The patient's legal guardians gave written informed consent for publication of this case report and accompanying images. The Editor-in-Chief of this journal keeps a copy of the written consent for review.

6.6.2. Conflicts of Interest

The authors declare no conflicts of competence or interest

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