

Clinical epidemiological retrospective study of glomus tumors diagnosed over 16 years in a reference unit

Estudo retrospectivo clinicoepidemiológico dos tumores glômicos diagnosticados ao longo de 16 anos em unidade de referência

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ABSTRACT

Introduction: *Glomus tumor is a benign and rare neoplasm that appears as solitary and painful nodule, mainly subungual. It presents characteristic clinical manifestations such as paroxysmal pain and sensitivity to local pressure and to cold.*

Objective: *To conduct a clinical epidemiological survey of glomus tumors diagnosed in a reference unit in 16 years.*

Methods: *Retrospective study assessing clinical and epidemiological data of 15 patients with glomus tumor confirmed by histopathological report from 2000 to 2016. Data on age, gender, tumor location, associated symptoms, duration of symptoms until diagnosis, clinical presentation, histological type and recurrence after surgery were analysed.*

Results: *Fifteen cases were diagnosed, 11 in women (73.3%) and four in men (26.7%). Mean age was 63 years. The most frequent location was the subungual region. Mean duration of symptoms until diagnosis was 8 years.*

Conclusions: *In this study, the number of cases of glomus tumor fluctuated throughout these 16 years, with a mean of 0.9 cases/year. A higher prevalence in women over 60 years, subungual region location and solid glomus histologic subtype was observed.*

Keywords: *glomus tumor; nail diseases; neoplasms*

RESUMO

Introdução: O tumor glômico é neoplasia benigna e rara que se apresenta como nódulo solitário e doloroso de localização principalmente subungueal. Apresenta manifestações clínicas características como dor paroxística e sensibilidade à pressão local e ao frio.

Objetivo: Realizar levantamento clinicoepidemiológico dos tumores glômicos diagnosticados em unidade de referência em 16 anos.

Métodos: Estudo retrospectivo analisando dados clínicos e epidemiológicos de 15 pacientes com diagnóstico de tumor glômico confirmado pelo laudo histopatológico no período de 2000 a 2016. Foram avaliados dados como idade, sexo, localização do tumor, sintomas associados, duração dos sintomas até o diagnóstico, apresentação clínica, tipo histológico e recidiva após a cirurgia.

Resultados: Foram diagnosticados 15 casos, em 11 mulheres (73,3%) e quatro homens (26,7%). A média de idade foi 63 anos. A localização mais frequente foi a região subungueal. O tempo médio da duração dos sintomas até o diagnóstico foi de oito anos.

Conclusões: Neste estudo, o número de casos de tumor glômico oscilou ao longo desses 16 anos, com média de 0,9 caso/ano. Observou-se maior prevalência em mulheres com mais de 60 anos, na região subungueal e do subtipo histológico glômico sólido.

Palavras-chave: tumor glômico; doenças da unha; neoplasia

INTRODUCTION

The glomus body is a highly specialized arteriovenous anastomosis responsible for thermoregulation.¹ It is located in the reticular dermis throughout the body, and is present in high concentrations in the digital extremities, especially underneath the nail.² As a result, the most common location of the tumor is the hands (75% of cases), with preference for the fingertips and subungual space. Other body sites, such as central nervous system, stomach, liver, mediastinum, trachea, lungs, bones, joints and genitals have been mentioned in the literature. Glomus tumor is a benign hamartoma of the glomus body,¹ corresponding to 1 - 4.5% of tumors of the hand.^{3,4} It was first described in 1812 by Wood as a very painful subcutaneous tumor with slow growth and sensitive to temperature variations.^{2,4} It emerges as an intensely sensitive (in special to cold), small, purple or bluish nodule, painful to the touch.^{1,4} Multiple locations are uncommon and mainly occur in children under 16 years old.² Solitary tumors are usually found in adults with usually between 30 and 50 years of age, with subungual location, predominantly in females.^{1,4} Solitary or multiple tumors have been described in association with neurofibromatosis type 1.¹ Congenital and hereditary forms have autosomal dominant inheritance.³ The tumor occurs by hyperplasia in one or more normal parts of the glomus body.² The pain develops from the contraction of glomus cells.² Clinical occurrences are hyperesthesia and/or localized paroxysmal pain, which increase with exposure to cold.² Hypersensitivity to cold is observed in 63 to 100% of cases located in the hands, however it is rarely found in extradigital tumors. The combination of magnetic resonance imaging, clinical tests and histopathology allows the diagnosis.² There are three main clinical tests for the diagnosis of this tumor.^{2,4} The first is the Love's pin test, when the area where the pain is located is gently pressed with the tip of a pin or clip; in subungual tumors the pressure is directed to the nail plate at different locations until the exact area of the sensitivity is determined.^{2,4} For a positive test, the patient should feel intense pain, sometimes with the jerking of the hand.² The Hildreth's test involves placing a tourniquet on the base of the finger where the tumor is located, subsequently applying the Love's test.^{2,4} For a positive result, the patient should not feel pain.^{2,4} The third test is that of the sensitivity to the cold, which is expected to produce increased pain with exposure to cold.²

Combined with the clinical tests, high-resolution imaging, such as magnetic resonance, has provided a valuable method for the diagnosis of the glomus tumor. Most of these tumors show high signal intensity on T2 and intense enhancement after gadolinium injection.² Plain radiography may evidence bone erosion, suggesting the diagnosis, nevertheless it is a rare finding and takes place in only 22% of cases.^{3,4} Other tests have been described, however with little contribution to the diagnosis.⁴

Histologic analysis of the glomus tumor characterizes it with the presence of small vessels with normal endothelium, circumscribed by compact nests of polygonal cells with round and central nuclei, and eosinophilic cytoplasm.⁵ Seven distinct histological forms are described. The glomangioma, which is the

most common variant, has a prominent vascular component, with the vessels' lumen being dilated or cavernous. Some cases can have hyalinization of the vessels' walls and thrombi within them. The glomangiomyoma is a rare type, being characterized by muscle cells increased in size and number, surrounding the vascular spaces. The other described types of glomus tumors are: solid, symplastic, glomangiomas, infiltrate and malignant.⁵ In most cases, surgical treatment leads to the complete resolution of symptoms.^{1,3} Recurrence after surgery occurs in 4 to 15% of cases, depending on the series.¹ Malignant transformation is rare (less than 1% of cases of glomus tumors) and occurs most commonly in non-acral locations.⁶

The objective of the present study was to perform a clinical epidemiological survey of glomus tumors diagnosed at a reference unit during a period of 16 years.

METHODS

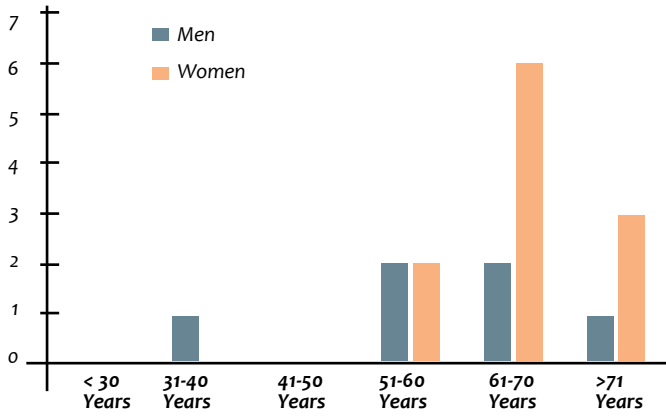
A retrospective review of medical records of patients diagnosed with glomus tumor was carried out at the Dermatology Division of the Instituto Lauro de Sousa Lima, in the city of Bauru (São Paulo, Brazil), covering a 16-year period (from 01/01/2000 to 01/01/2016). The following variables were analyzed: age, gender, tumor location, associated symptoms, duration of symptoms up until diagnosis, clinical appearance, histological type and recurrence after surgery.

RESULTS

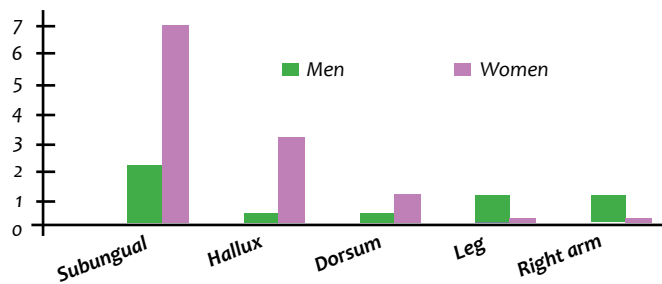
Between the years of 2000 and 2016, 15 cases of glomus tumor were diagnosed (11 women = 73.3% and 4 men = 26.7%, all Caucasian) (Graph 1). The average age at diagnosis was 63 years (min = 36, max = 85), with a predominance of the 61 to 70 years old age group (40%). The average age of women was slightly higher than that of men (66 and 54, respectively). Regarding the location of the lesion, the most common topography was the subungual in the fingers (60%), followed by the hallux (20%), dorsum (6.8%), lower limbs (6.7%) and arms (6.7%). In both genders the glomus tumor occurred preferably in the subungual region (men = 50%, women = 73%) (Graph 2). The average time of onset of symptoms at diagnosis was 8 years (min = 1, max = 30) (Graph 3). Clinical abnormalities most frequently found were: erythematous-violet dotted area visualized through the nail plate (40%); onychodystrophy (26.7%); medial fissure in the nail plate (20%); and painful bluish nodule in the skin (13.3%) (Graph 4). Pain was present in 100% of patients. The most common histological type was the solid glomus tumor (86.7%), followed by glomangiomas (13.3%). Regarding the treatment, all patients underwent surgical excision, with 13.3% presenting recurrence.

DISCUSSION

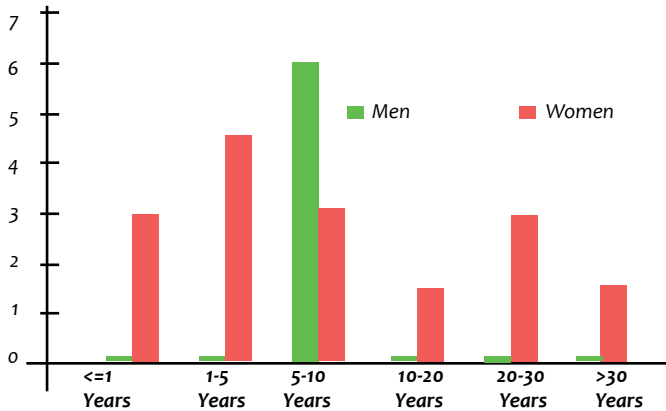
In the present study, the number of diagnosed cases of glomus tumor fluctuated throughout the study period, with a mean value of 0.9 case/year. Regarding the topography, the most common site was the subungual region of the upper limbs



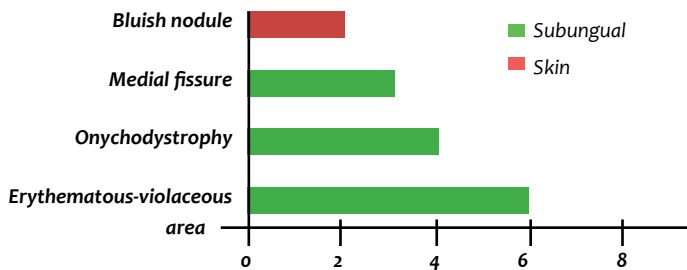
GRAPH 1: Number of glomus tumor cases distributed by gender and age



GRAPH 2: Location of glomus tumor by gender



GRAPH 3: Time of onset of symptoms at diagnosis of the glomus tumor by gender.



GRAPH 4: Clinical manifestations of the glomus tumor by location

(60%), followed by the hallux (20%), dorsum (6.8%), legs (6.7%) and arms (6.7%). The average time to diagnosis was 8 years. Unlike the present study, a review performed by the Mayo Clinic with 56 extradigital cases evidenced prevalence in the forearms (20%) and knees (18%).⁷

A higher prevalence of glomus tumor was observed in women over 60 years of age and in the subungual region of the upper limbs. These data are in line with the literature, nonetheless differing in age, with the present study evidencing cases at an older age group.^{3,4}

The most frequently found histological subtype was the solid glomus tumor (86.7%), a fact that did not corroborate findings of previous studies, which suggest the glomangioma as the most frequent.⁵ The most often observed clinical appearance of the subungual glomus tumor was an erythematous-violaceous area (40%). The clinical tests, useful due to their easy implementation and cost effectiveness, have been performed only in 2 of the 15 cases (13.3%), yielding positive results in both.

Imaging studies were performed in 6 cases (40%), with plain radiography being performed in 4 of them. In only 1 case focal osteolysis was observed in the distal phalanx (25%). Magnetic resonance imaging was performed in 1 case, evidencing the tumor and its topography, leaving no doubt as to the diagnosis. Ultrasonography was requested in 1 case, however it did not evidence injury.

The found recurrence rate was 13.3%, coinciding with that in the literature.^{3,4}

CONCLUSIONS

The present study has demonstrated that the glomus tumor is associated to a delay in its diagnosis, leading the patient to live with chronic pain and which most often has excellent response to surgical treatment. Although this entity is more often found in the subungual region, it can emerge in other body sites. In addition, specific clinical tests are excellent, cost-effective tools when there is suspicion of the diagnosis. ●

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