# **GLOMUS TUMORS**

W. DE MAERTELEIRE, P. NAETENS, L. DE SMET\*

The authors report a series of 12 consecutive patients with 13 glomus tumors operated from July 1991 until February 1999. Symptoms were present for an average of 1.9 years before surgery. Women were more frequently affected. The mean age was 44 years. In 12 of the 13, the tumor was located in the distal phalanx and one patient had a glomus tumor on the dorsum of the hand. One glomus tumor was found in the right hallux of a two-year-old child. Both hands and all fingers were equally involved. One bilateral glomus tumor was associated with neurofibromatosis. All tumors were resected and histology confirmed the diagnosis. The result was good with immediate pain relief. No recurrence has been noted to date.

**Keywords:** hand, glomus tumor. **Mots-clés:** main, tumeur glomique.

#### INTRODUCTION

The glomus tumor was first described by Masson in 1924. It is a benign tumor which develops from the neuromyoarterial elements of the glomus bodies. The etiology is unknown. The most common location for the glomus tumor is the distal phalanx (13). It is a small tumor which is frequently subungual. It is usually observed only by a slight discoloration of the skin or the nail. This discoloration may sometimes be enhanced by transillumination (4). Symptoms consist of the classical triade: pain, cold intolerance and tenderness. In most cases the pain and tenderness are very defined. When the tumor is located in the nail bed, a nail deformity can be seen. The tumor is frequently not recognized for many years. Most patients are symptomatic a long time before effective treatment. Technical investigations are rather nonspecific. Radiography sometimes shows a small impression or sclerotic reaction in the bone adjacent to the tumor. Therefore it is important to compare normal and symptomatic sides. Ultrasonography is more specific and it should allow differentiation from ganglia or cysts. Scintigraphy is said to be positive in all cases but is nonspecific. Arteriography causes more frequent morbidity compared to other investigational methods. High resolution MRI is a very sensitive and specific but expensive investigation (2, 3). The only effective treatment is excision. The histopathology is well recognized. Histologically the tumor consists of well differentiated clusters of small polygonal cells with dark round nuclei and scanty cytoplasm around small blood vessels.

#### MATERIAL AND METHODS

Here we present 13 consecutive glomus tumors in 12 patients (8 women and 4 men) operated between July 1991 and February 1999. The ages ranged from 2 to 69 years with an average of 44.75 years. One case was bilateral; 5 cases involved the left hand and 5 cases the right hand. In one case the right hallux was involved. In all but one case the tumor was situated distal to the distal interphalangeal joint. In one case the tumor was situated dorsally to the base of the fifth metacarpal. In six cases the tumor was subungual and in the remaining six cases the tumor was located in the pulpa. The size of the tumor ranged from a few millimeters, no larger than a rice grain, to 1.5 cm in diameter. The largest tumor was

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found on the dorsum of the hand over the base of the fifth metacarpal. This tumor was easily palpable as a renitent mass. In this series, 7 cases had a noticeable discoloration of the skin or the nail (fig. 1). All patients had pain and tenderness but only one patient suffered from nocturnal pain. Time between onset of symptoms and diagnosis ranged from 6 months to 3 years with an average of 2 years. Cold intolerance was present in five cases. Xrays were made in 11 of the 13 cases, but only five xrays showed some periosteal impression or sclerosis (fig. 2). Three ultrasound examinations were done, of which only two showed a soft tissue tumor. One CT-scan and one technetium scintigraphy were performed, both negative. In 8 cases the diagnosis could be made by history and physical examination. In one case the preoperative tentative diagnosis was a foreign body, in two cases a preoperative diagnosis of neurofibroma was proposed and in another two cases a hemangioma was suspected. The diagnosis was confirmed histologically in all cases. In 7 cases surgery was performed under local anesthesia, in 4 cases under Bier's block and in one case (a twoyear-old child) under general anesthesia. For pulpar lesion the incision was straight over the tender spot (fig. 3a). For subungual lesions an incision on the lateral border was used (fig. 3b). Nail and nail bed were avoided with surgery as much as possible. However, the nail plate had to be incised and the nail itself partially or totally removed in 5 cases.

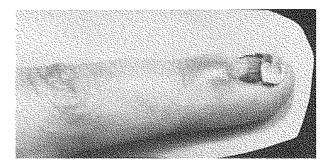


Fig. 1. — Clinical aspect of a glomus tumor.

### RESULTS

Follow-up in this study ranged from 7 months to 8 years and 1 month with an average of 2 years and 9 months. All patients had complete relief of pain postoperatively. No recurrences were reported and all diagnoses were confirmed histologically. Postoperatively one patients, who had neurofibromatosis, complained of a neurinoma-like tenderness in the scar. Two patients, in which the nail had to be partially removed during surgery, had a minor nail deformity. All other scars healed perfectly.

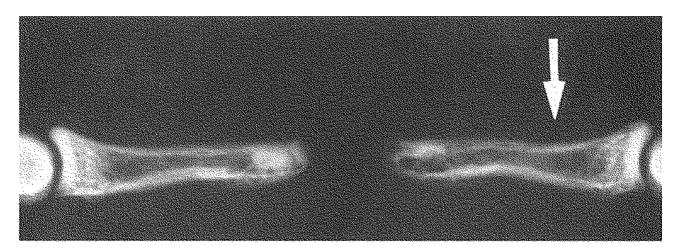
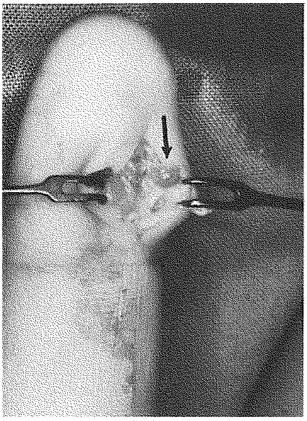
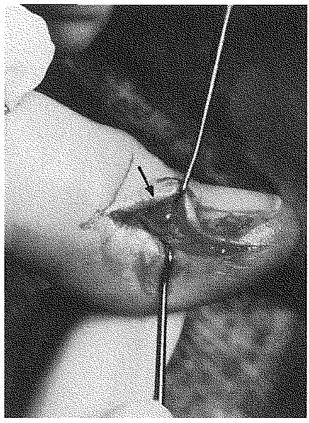


Fig. 2. — Radiological aspect with compression of the dorsal cortex of the terminal phalanx.

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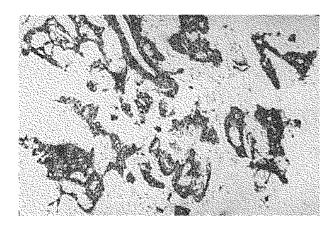
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Fig. 3. — Intra-operative aspect of a pulpar (a) and subungual (b) glomus tumor.

### DISCUSSION

The glomus tumor is a benign tumor usually localized in the distal phalanges of the hand, preferentially in the subungual tissues. These features were also found in this series. As in many published series we found the lesion occurred preferentially in middle-aged women. One patient, a 52-year-old woman, presented with a bilateral solitary glomus tumor; both tumors were located in the pulpa. This patient also had neurofibromatosis. In 1995, Sawada et al. described three cases of neurofibromatosis and glomus tumor (12). Glomus tumors are not familial; they usually occur in places were glomus bodies are located. These places are regions of extensive arteriovenous shunting, although some glomus tumors have been described in aberrant places where such arteriovenous shunting is absent. Although many believe that glomus tumors originate from smooth muscle cells (10, 11), Kline *et al.* published a case report in which substantial evidence was found for a glomus tumor originating from a digital nerve (8). Others also subscribe to the hypothesis of a neural crest cell origin (14). It is striking to see that our patient with a disorder of the nervous system was the only patient with simultaneous bilateral glomus tumors. In the histologic specimens of this patient, prominent neural tissue could be found within the glomus tumors, so we can confirm that there is some evidence for a neural origin or at least a relationship between the nervous system and some glomus tumors.

One tumor was found in a two-year-old child. This child presented with a nail deformity of the right hallux and refused to wear shoes that put



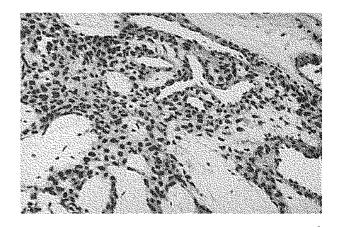


Fig. 4. — Histological aspect.

pressure on the nail plate. The mother stated that there had always been a nail deformity since birth. Congenital glomus tumors have been described but were mostly located on the abdomen as indurated painless plaques (6, 9). In the middle ear and skull base congenital glomus tumors have been reported, but to our knowledge no congenital glomus tumor in the hand or foot has been described before (7). One case of a congenital glomus tumor in the patella has been described (1).

The fact that it is a rare tumor constituting only about 2% of all tumors found in the hand, may explain the long delay before the right diagnosis is made and therapy is established (13). The preoperative diagnosis can be made from the history and the clinical examination. Love's test (1944), which consists of applying localized pressure with the head of a straight pin, can help to make the diagnosis and to localize the tumor precisely (referred in 13). Cold intolerance is a specific sign but rather difficult to test in an outpatient clinic. The ischemia test described by Hildreth is another rather specific test. In this test an arm cuff is inflated well above the arterial pressure. Owing to ischemia of the tumor, tenderness and pain disappear (5).

Technical investigations are of little value since ultrasound and plain xrays prove negative or non-specific. Scintigraphy should be positive in all cases, but proved nonspecific. MRI was never done in this series.

The only known cure is surgical excision. As the tumor is usually very small and located in the hand, the procedure can usually be performed under local anesthesia. In our series only one case was treated under general anesthesia because the patient was a 2-year-old child. In 4 cases anesthesia was done intravenously with marcaine. If the tumor is subungual, a lateral approach is used whenever possible. This lateral approach provided good scar healing and no nail deformity while the transungual approach gave a nail deformity in two of the five cases.

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#### SAMENVATTING

W. DE MAERTELEIRE, P. NAETENS, L. DE SMET. Glomus tumoren.

De auteurs beschrijven een reeks van 12 patiënten met 13 glomus tumoren (12 in de hand en één in een teen) operatief behandeld tussen 1991 en 1999. De symptomen bestonden gemiddeld twee jaar preoperatief. Er was een voorkeur voor vrouwen met een gemiddelde leeftijd van 44 jaar. Bij 12 gevallen was er een lokalisatie in de

distale phalanx, éénmaal t.h.v. de dorsâle zijde van de metacarpaal. Er was geen verschil tussen links en rechts en evenmin tussen pulpaire of subunguale lokalisatie. Alle stralen waren evenveel aangetast. Een bilaterale lokalisatie zag men bij één patient, met neurofibromatose, éénmaal was er en lokalisatie in de teen bij een tweejarig kind. In alle gevallen bestond uit resectie en werd de diagnose histologisch bevestigd. Het resultaat was goed in alle gevallen met onmiddellijk verdwijnen der klachten en geen enkel recidief werd tot heden vastgesteld.

## RÉSUMÉ

W. DE MAERTELEIRE, P. NAETENS, L. DE SMET. Les tumeurs glomiques.

Les auteurs présentent une série de 12 patients porteurs de 13 tumeurs glomiques (12 au niveau de la main, 1 au niveau du pied), opérées entre juillet 1991 et février 1999. Les symptômes étaient apparus en moyenne un peu moins de deux ans avant l'opération. Il y avait une prédominance du sexe féminin ; l'âge moyen était de 44 ans. Douze fois sur treize, la tumeur était localisée au niveau de la phalange distale, un patient avait une tumeur glomique à la face dorsale d'un métacarpien. En outre, les auteurs ont traité une tumeur glomique du gros orteil chez un enfant de deux ans. Il n'y avait pas de prédominance gauche ou droite; tous les rayons étaient également affectés. Une tumeur glomique bilatérale était associée à une neurofibromatose. Dans tous les cas, la tumeur a été réséquée, le diagnostic a été confirmé histologiquement. Le résultat a été bon, avec soulagement immédiat des douleurs. Aucune récidive n'a été notée à ce jour.