

Atypical presentation and management of a subungual glomus tumor in the pediatric population: a case report

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The glomus tumor is a rare vascular neoplasm that accounts for less than 2% of all soft tissue tumors of the hand. This is the first case study to describe the presentation and curative resection of a subungual glomus tumor in the pediatric population. The glomus tumor is classically described as a solitary subungual mass in women aged 30–50 who present with the triad of pain, temperature sensitivity, and point tenderness. However, given an infrequent and inconsistent presentation, the typical time from onset of symptoms to the correct diagnosis is 7 years, resulting in significant functional and cosmetic morbidity for patients. Here we describe an unusual case of a 6-year-old boy who presented with 3 years of pain and progressive, nontraumatic deformity of his left thumb nail. The patient underwent a left thumb nail plate removal, excision of the subungual mass, and nail

bed repair. The diagnosis of subungual glomus tumor was confirmed on pathology. Glomus tumors are rare in the pediatric population but should be considered in patients presenting with nail pain and deformity. *Ann Pediatr Surg* 14:89–91 © 2018 Annals of Pediatric Surgery.

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Introduction

Glomus tumors are rare, benign vascular proliferations, accounting for 2% of all hand tumors [1]. Diagnosis and management has not been reported in the pediatric population. The most common presentation is a firm, solitary paraungual or subungual nodule in the distal phalanx that results in moderate pain and point tenderness. Because the mass is often too small to be identified on physical examination, it is typically several years before a correct diagnosis is made and curative surgical excision is achieved. Delay in diagnosis or misdiagnosis may be even more common in the pediatric patient because the diagnosis is even more rare than in the adult population and often is not included in the differential diagnosis of a painful digital mass in children. Here we describe a 6-year-old boy who presented with a solitary subungual glomus tumor that was successfully surgically excised.

Case report

The patient was a 6-year-old right hand dominant boy who presented with a painful, progressive, nontraumatic deformity of his left thumb nail. The patient's mother reported a 3-year history of significant tenderness of the left thumb upon contact or compression. He had been seen by his pediatrician and several consultants with no definitive diagnosis. The patient described experiencing pain in the thumb multiple times per day whenever pressure was applied to the tip of his thumb. The patient did not describe night pain or cold sensitivity, and there was no history of extremity injury, trauma, or systemic complaints.

The physical exam revealed a mass deep to the nail plate in the central third of the proximal nail as shown in Fig. 1.

In addition, the overlying sterile matrix was deformed with secondary nail deformity resulting in a split at the junction of the radial and middle thirds of the nail. The radial aspect of the nail was delaminated from the sterile matrix, resulting in a foreshortened nail plate in this area. Pain was elicited by palpation and compression. A three-view

Fig. 1



Dorsal image of the left thumb showing nail deformity and mass subungual mass.

Fig. 2

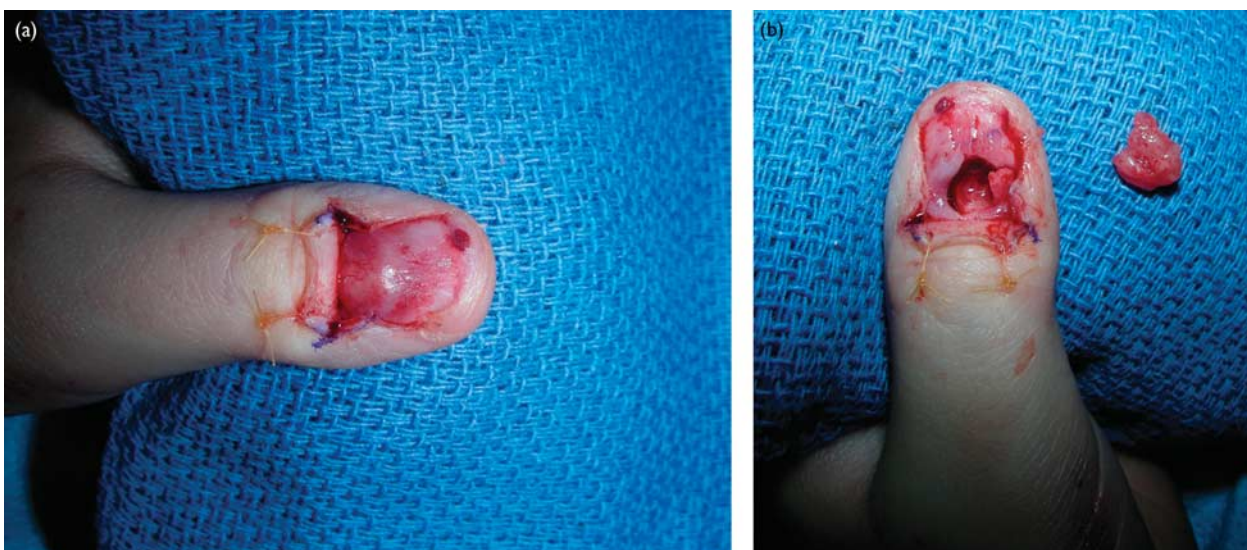


Preoperative lateral radiograph of the left thumb revealed indentation of the dorsal cortex of the distal phalanx.

radiograph of the left thumb revealed indentation of the dorsal cortex of the distal phalanx (Fig. 2).

The patient underwent an excision in which the nail bed was incised, elevated off of the glomus tumor, and retracted. The glomus tumor was resected and the nail bed was repaired with absorbable suture, covering the indentation in the bone, as shown in Fig. 3. The resulting

Fig. 3



Operative images display (a) left thumb with nail plate removed and (b) after excision of tumor.

Fig. 4



At 1-year follow-up, the patient's nail revealed regrowth of a normal nail. The patient also reported resolution of preoperative symptoms of pain and tenderness.

histopathology revealed a tumor composed of small, uniform, and rounded cells with centrally placed round nuclei, forming in nests, consistent with a glomus tumor.

At 1-year follow-up, the patient reported resolution of preoperative symptoms of pain and tenderness and had regrowth of a normal nail (Fig. 4).

Informed written consent was obtained from the patient's parent for the surgery and publication of the photographs for this study.

Discussion

Glomus tumors are benign vascular proliferations of the neuromyoarterial apparatus involved in the thermoregulation of skin circulation. They arise from glomus bodies, which are found in the dermal reticular layer of the skin and are highly concentrated in the finger tips, particularly beneath the nail bed [2]. This neoplasm can occur anywhere in the skin or soft tissue, although the most common presentation is a firm, solitary nodule in the distal phalanx in a paraungual or subungual location.

Glomus tumors are rare, accounting for 2% of all hand tumors [1]. The average age of presentation is between age 30 and 50, and the tumor is three times more frequent in female individuals [3]. The tumor is particularly rare in pediatric patients. Colon and Upton [4] reported a series of 349 pediatric hand tumors, nine of which were glomus tumors, located within the digits or thumb. Of those, the two youngest patients were also 6 years of age but none of the cases were described (personal communication, Lonnie Perkins, 4 September 2015). There have been no other pediatric cases of glomus tumors reported in the English literature.

Although patients with glomus tumor seek medical attention early for moderate pain and point tenderness, the mass is often too small to be identified on physical exam and it is typically several years before a correct diagnosis is made. The presence of pain often leads to misdiagnoses such as neuroma or gouty arthritis [5]. The classic triad of paroxysmal pain, pinpoint tenderness, and cold hypersensitivity is inconsistently present. Diagnosis was delayed in this particular case in part due to the patient's young age and atypical presentation lacking night-time pain and cold intolerance.

Specific clinical tests such as the Love's sign, Hildreth's test, and cold sensitivity can aid in the diagnosis. Love's sign, in which pressure applied over the lesion with the end of a pencil elicits severe pain, was found to be 100% sensitive and 78% accurate. A positive Hildreth's test occurs when slow insufflation of a brachial cuff diminishes the point pain elicited in the Love's sign. This test was found to be 71.4% sensitive and 100% specific. A cold

sensitivity test has been reported to be 100% sensitive and specific [6]. These tests were not performed in the case described, as the glomus tumor was not on the differential list at the time of examination given the atypical presentation. The typical differential symptoms for the pediatric patient with subungual mass include blue nevus, venous malformation, and schwannoma.

The use of MRI has been reported as a useful tool to confirm tumor location [2]. MRI may show a high signal on T2 and low signal on T1 [7]. In long-standing cases, bony erosions may be evident on plain radiography.

The standard treatment for glomus tumors is complete surgical excision of the mass and biopsy. Patients typically experience a complete resolution of symptoms. Risk of recurrence increases with incomplete excision. If symptoms such as pain and pinpoint tenderness persist for greater than 3 months, repeat imaging and re-exploration of the area is recommended [8].

In summary, we identified an unusual case of subungual glomus tumor in a pediatric patient. Given an atypical presentation, diagnosis and curative resection were delayed. The patient was successfully treated with surgical excision of the palpable mass with resolution of symptoms. Awareness that atypical presentation of glomus tumors can be a cause of hand pain in children is essential for early diagnosis and treatment of this rare, but painful condition.

Conflicts of interest

There are no conflicts of interest.

References

- 1 Tang CYK, Tipoe T, Fung B. Where is the lesion? Glomus tumours of the hand. *Arch Plast Surg* 2013; **40**:492–495.
- 2 McDermott EM, Weiss A-PC. Glomus tumors. *J Hand Surg [Am]* 2006; **31**:1397–1400.
- 3 Lee CH, Tandon A. Focal hand lesions: review and radiological approach. *Insights Imaging* 2014; **5**:301–319.
- 4 Colon F, Upton J. Pediatric hand tumors. A review of 349 cases. *Hand Clin* 1995; **11**:223–243.
- 5 Hazani R, Houle JM, Kasdan ML, Wilhelmi BJ. Glomus tumors of the hand. *Eplasty* 2008; **8**:e48.
- 6 Bhaskaranand K, Navadgi BC. Glomus tumour of the hand. *J Hand Surg* 2002; **27B**:229–231.
- 7 Kim DH. Glomus tumor of the finger tip and MRI appearance. *Iowa Orthop J* 1999; **19**:136–138.
- 8 Rao RVMS, Raja DA. Three cases of subungual glomus tumours of the fingers – a case series. *J Clin Diagn Res* 2014; **8**:ND05–ND06.