Primary Intraosseous Glomus Tumor: A Case Report

JONGKOLNEE SETTAKORN, M.D.*,
PREECHA CHALIDAPONG, M.D.**,
SAMREUNG RANGDAENG, M.D.*
OLARN ARPORNCHAYANON, M.D.**,
BENJAPORN CHAIWUN, M.D.*,

Abstract

We reported a rare case of a primary intraosseous glomus tumor in a 53-year-old woman who had a small lytic lesion in the distal phalanx of her left index finger. The radiologic appearance showed a well circumscribed osteolytic lesion without sclerotic rim. Histologic examination revealed solid nests or sheets of polygonal cells surrounding branching vasculatures consistent with a glomus tumor. Curettage resulted in complete removal of the tumor as well as pain alleviation. The patient was well without evidence of recurrent disease following an 18 month follow-up. Despite its rarity, intraosseous glomus tumor should be included in the differential diagnosis of bone lesions in which plain radiography showed a well circumscribed punch-out lesion without sclerotic rim especially those arising in the finger.

Key word : Intraosseous Glomus Tumor, Glomus Tumor, Pericyte

SETTAKORN J, ARPORNCHAYANON O, CHALIDAPONG P, CHAIWUN B, RANGDAENG S
J Med Assoc Thai 2001; 84: 1641-1645

The glomus tumor, first described by Masson in 1924[1], is a benign lesion that is a neurally innervated arteriovenous anastomosis invested by specialized muscle cells known as glomus cells. These tumors arise most frequently in the subungual region of the fingers. Less than 100 cases of glomus tumor involving bone[2] have been reported in the literature following the first description by Iglesias et al 1939 [3]. Most cases are likely caused by direct extension from an adjacent subcutaneous site. Approximately 18 cases of glomus tumors arising intraosseously have been previously mentioned[1-14] in the literature. We present an additional case with its characteristic clinical and microscopic features.

* Department of Pathology,
** Department of Orthopedics, Faculty of Medicine, Chiang Mai University, Chiang Mai, 50200, Thailand.

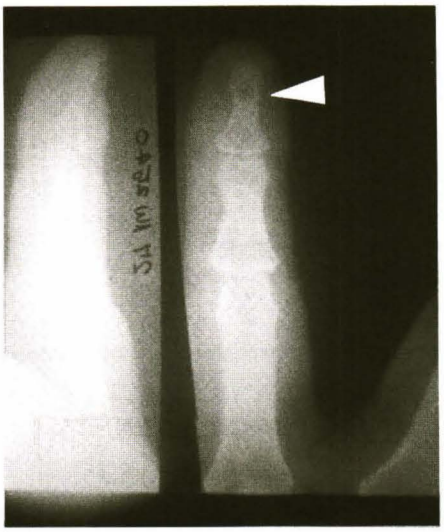

Fig. 1. **Plain radiography of subject's left index finger showed a small, punch-out, radiolucent lesion without sclerotic rim (Arrow head).**

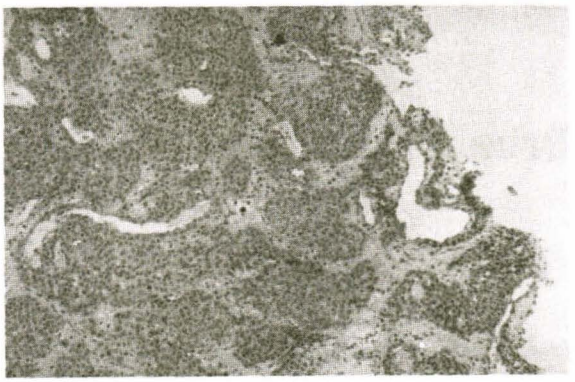

Fig. 2. **Microscopically, the tumor is composed of broad sheets and solid nests of uniform polygonal cells surrounding branched thin wall blood vessels. (H&E, 100X).**

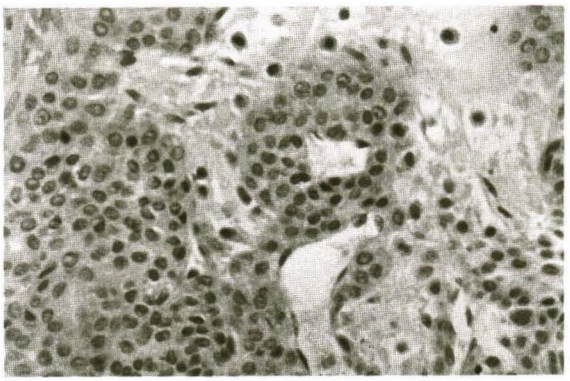

Fig. 3. **On higher power, the tumor cells are of uniform polygonal shape with round to oval, centrally placed vesicular nuclei and small nucleoli. The cytoplasmic appearances varied from pale to solid eosinophilic. (H&E, 400X).**

A CASE REPORT

A 53-year-old woman came to the hospital with a 3 year-history of localized stabbing pain in her left index finger. There was no previous known history of injury or infection at that location. On physical examination, there was a mild swelling with a point of tenderness at the mid pulp of the left index finger. No skin discoloration was observed.

Radiographic appearance of the distal phalanx showed a well circumscribed, oval, radiolucent area in the distal half without peripheral sclerosis (Fig. 1). The bony cortex was intact. Blood hematology, chemistry and serological examination were normal. The preoperative diagnosis included enchondroma, glomus tumor, dermoid cyst and giant cell tumor of the bone.

Pathologic Findings

The operation with hockey stick incision and total tumor removal was performed under regional anesthesia. The tumor was red brown, well circumscribed with soft consistency, measuring 3 mm in diameter.

Microscopically, the tumor was composed of branching vascular spaces surrounded by broad sheets or nests of glomus cells (Fig. 2). The vascular spaces were lined by a single layer of endothelial cells. The glomus cells were of polygonal shape with an indistinct cellular border containing pale eosinophilic cytoplasm (Fig. 3). Their nuclei were uniform, round with inconspicuous nucleoli.

Periodic acid Schiff's stain failed to demonstrate intracytoplasmic glycogen.

Immunohistochemical studies using antibodies directed against smooth muscle actin (clone 1A4, Dako Corporation, Glostrup, Denmark) and against vimentin (clone V9, Dako Corp.) showed diffuse cytoplasmic immunoreactivity with both markers.

One year after operation, the patient was well without evidence of recurrent disease.

Comment

There were three patterns in which glomus tumors involved the bone that could be distinguished radiographically[2]. The first pattern was overlying glomus tumors of the adjacent soft tissue that pinched onto the bone without direct bony invasion, this pattern resulted in deformity of the bony cortex. The second pattern represented glomus tumors that could erode bony cortex and became intraosseous tumor by direct extension. Radiologic findings of this pattern showed a well-demarcated, eccentric bony defect that extended through the cortex into the intramedullary canal.

The third pattern of skeletal involvement of glomus tumors were those arising intraosseously and radiologically, produced a "punched out" lytic area surrounded by intact cortex. This pattern of skeletal involvement was the rarest form among intraosseous glomus tumor, hence, to the best of our knowledge, only 18 cases of primary intraosseous glomus tumor have been reported in the world literature[1-14].

Pathogenesis of intraosseous glomus tumor was thought to be the result of the presence of normal glomus organelles in the medullary cavity and the occurrence of the tumor during differentiation of multipotential mesenchymal stem cell[3]. Their locations and clinical information are summarized in Table 1. These tumors were rarely found in children and adolescents. There was only a single case of a 14-year-old girl who had the tumor at the coccyx [10]. The usual age range was 14-61 years-old with a slight female predominance.

The most common site of the tumor was the distal phalanx of the finger. The left hand was more frequently encountered than the right side in which the thumb and little finger were the two most common sites. The other sites included the base of the middle phalanx[5], ulnar[6], thoracic spine[7], lumbar spine[8], sacrum[9] and coccyx[10].

The tumor was usually small ranging from a few millimeters up to 2 cm. The major complaint of patients with this tumor was stabbing pain at the affected sites with pain duration varying from 10 months to 5 years.

Table 1. Summary of 18 reported cases of intraosseous glomus tumor.

Case No.	Authors (year)	Age	Sex	Location
1	Iglesias et al (1939)	32	Female	Distal phalanx – Lt ring finger
2	Lattes and Bull (1948)	28	Female	Distal phalanx – Rt thumb
3	Leshman and Kraissl (1949)	48	Female	Distal phalanx - Lt little finger
4	Lashman and Kraissl (1949)	36	Female	Distal phalanx - Rt thumb
5	Machenzie (1962)	30	Female	Distal phalanx - Lt little finger
6	Mackenzie (1962)	61	Female	Distal phalanx - Lt thumb
7	Siegel (1967)	24	Male	Distal phalanx - Lt index finger
8	Ishii et al (1973)	60	Male	Distal phalanx - Lt little finger
9	Sugiura (1976)	33	Female	Distal phalanx - Lt little finger
10	Ho and Pak (1980)	27	Female	Coccyx
11	Pambakian and Smith (1981)	14	Female	Coccyx
12	Pambakian and smith (1981)	35	Female	Coccyx
13	Bjorkengren et al (1986)	68	Male	Middle phalanx - Rt little finger
14	Rozmaryn et al (1987)	24	Female	Rt proximal ulnar
15	Kobayashi et al (1990)	22	Female	Sacrum
16	Bessho et al (1991)	49	Male	Thoracic spine (T2 body, Lt)
17	Simmons et al (1992)	30	Male	Distal phalanx - Lt thumb
18	Robinson et al (1996)	45	Female	Lumbar spine (L1 pedicle, Rt)
19	Present report	53	Female	Distal phalanx - Lt index finger

Rt = right, Lt = left

Radiologically, a glomus tumor produced a well defined, oval, lytic lesion resembling enchondroma, epidermal inclusion cyst, unicameral bone cyst, sarcoidosis, tuberous sclerosis and xanthoma of bone[9]. The other differential diagnoses included oseoid osteoma, subungual melanoma, metastatic carcinoma and osteomyelitis[1].

The key histologic feature[1] for diagnosis was the demonstration of nests or masses of glomus cells surrounding thin wall vessels. The tumor cells bore round to oval nuclei set in a light to dense granular eosinophilic cytoplasm. The stroma comprised either fibrous or fibromyxoid material. Anaplasia was invariably absent. Mitoses were rare. Under low power magnification, the tumor might superficially resemble cavernous hemangioma, hemangiopericytoma or paraganglioma. In doubtful cases, reticulin stain might be helpful by revealing conformed reticulin framework wrapped around individual tumor cells. Intracytoplasmic glycogen was characteristically absent.

Immunohistochemically, vimentin and muscle actin could be demonstrated in nearly all tumors as shown in our case while desmin might give inconsistent results. Laminin and factor IV collagen had been shown to outline individual cells as well as tumor cell nests[10].

By electron microscopy[1], the glomus cells were surrounded by a thick basal lamina. Their cytoplasm contained abundant thin filament, pinocytic vesicles, mitochondria and endoplasmic reticulum. These features suggested their smooth muscle rather than pericytes origin.

The treatment of choice was total excision. Nonetheless, in case of a small tumor in the distal phalanx, curettage was considered sufficient; and consequently, almost all cases were treated by curettage or total excision of the tumors. Amputation was performed in only two reported cases[4]. Natural history of this tumor was uniformly benign and no recurrent case was encountered in the literature.

In summary, we reported an additional case of primary intraosseous glomus tumor with characteristic clinical, radiologic and pathological presentations demonstrated. Although this tumor is extremely rare, an intraosseous glomus tumor should be included in the differential diagnosis of bone lesions in which plain radiography showed a well circumscribed punch-out lesion without sclerotic rim especially those arising in the finger.

(Received for publication on April 21, 2001)

REFERENCES

1. Mirra JM. Vascular tumors. In: Mirra JM, editor. Bone tumors. Philadelphia: Lea & Febiger, 1989: 1335-479.
2. Fechner RE, Mills SE. Tumors of the bones and joints. In: Rosai J, Sobin LH, editors. Atlas of tumor pathology 3rd series. 8th fascicle. Washington DC: AFIP, 1993: 129-44.
3. Iglesias de la Torre L, Gomez Camejo M, Palacios G. Consideraciones clinicas, anatomicas, radiologicas y quirurgicas del glomus tumoral de Masson. Cirurgia Ortopedica y Traumatologia (Habana) 1939; 7: 11-7.
4. Sugiura I. Intra-osseous glomus tumour. J Bone Joint Surg (Br) 1976; 58: 245-7.
5. Bjorkengren AG, Resnick D, Haghighi P, Sartoris J. Intraosseous glomus tumor: Report of a case and review of the literature. Am J Orthop 1986; 147: 739-41.
6. Rozmaryn LM, Sadler AH, Dorfman HD. Intraosseous glomus tumor in the ulnar. A case report. Clin Orthop 1987; 220: 126-9.
7. Bessho Y, Kataoka O, Sho T, Kitazawa S, Okada S. Intraosseous glomus tumor in the upper thoracic spine complicating compression myelopathy. Spine 1991; 16: 989-90.
8. Robinson JC, Kilpatrick SE, Kelly DL JR. Intraosseous glomus tumor of the spine: Case report and review of the literature. J Neurosurg 1996; 85: 344-7.
9. Kobayashi Y, Kawaguchi T, Imoto K, Yamamoto T. Intraosseous glomus tumor in the sacrum. A case report. Acta Pathol Japan 1990; 40: 856-9.
10. Pambakian A, Smith MA. Glomus tumours of the coccygeal body associated with coccydynia. A preliminary report. J Bone Joint Surg 1981; 63: 424-6.
11. Huvos AG. Bone tumor: Diagnosis, treatment and

prognosis. 2nd edition. Philadelphia: WB. Saunders, 1991: 553-74.
12. Enzinger FM, Weiss SW. Soft tissue tumor. 3rd ed. St. Louis: Mosby-Year book, 1995: 701-33.
13. Simmons TJ, Bassler TJ, Schwinn CP, Forrester DM. Primary glomus tumor of bone. Skeletal Radial 1992; 21: 407-9.
14. Sunderraj S, al-Khalifa AA, Pal AK, Pim HP, Sabri SH. Primary intraosseous glomus tumour. Histopathology 1989; 14: 532-6.

โกลมัส ทูเมอร์ ในเนื้อกระดูก

จงกลณี เศรษฐกร, พ.บ., โอฬาร อาภรณ์ชยานนท์, พ.บ.**,*
*ปรีชา ชลิดาพงศ์, พ.บ.**, เบญจพร ไชยวรรณ์, พ.บ.*, สำเริง รางแดง, พ.บ.**

รายงานผู้ป่วยโรค glomus tumor ที่เกิดขึ้นในเนื้อกระดูก จำนวน 1 ราย โดยเป็นผู้ป่วยหญิงไทยอายุ 53 ปี มีรอยโรคแบบ lytic lesion ที่ ส่วนปลายสุดของกระดูก distal phalanx ของนิ้วก้อยของมือซ้าย จากภาพถ่ายทางรังสีพบว่ารอยโรคมีขอบเขตชัดเจนและไม่พบลักษณะ sclerotic rim เมื่อนำชิ้นเนื้อมาดูด้วยกล้องจุลทรรศน์ พบกลุ่มของเซลล์ล้อมรอบหลอดเลือดที่แตกแขนง ซึ่งเข้าได้กับ glomus tumor หลังจากผ่าตัดเอาเนื้องอกออก อาการเจ็บของผู้ป่วยทุเลาลง ในผู้ป่วยที่มีอาการเจ็บที่ปลายนิ้วและมีรอยโรคแบบ lytic ในเนื้อกระดูก ควรคิดถึง glomus tumor ไว้ในการวินิจฉัยแยกโรคด้วย

คำสำคัญ : โกลมัส ทูเมอร์ ในเนื้อกระดูก, โกลมัส ทูเมอร์, pericyte

จงกลณี เศรษฐกร, โอฬาร อาภรณ์ชยานนท์,
ปรีชา ชลิดาพงศ์, เบญจพร ไชยวรรณ์, สำเริง รางแดง
จดหมายเหตุทางแพทย์ ฯ 2544; 84: 1641-1645

* ภาควิชาพยาธิวิทยา,
** ภาควิชาออร์โธปิดิกส์, คณะแพทยศาสตร์ มหาวิทยาลัยเชียงใหม่, เชียงใหม่ 50200