Intraosseous Proliferative Sparganosis : A Case Report and Review of the Literature

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Abstract

Intraosseous proliferative sparganosis is an extremely rare parasitic disease in which the larvae of incomplete differentiated sparganum proliferate in the human bone. We present the first case of intraosseous proliferative sparganosis arising in the long bone. The patient was a 51-year-old man who complained of a slow growing painful mass on his right leg. The radiographic findings showed an infiltrative osteolytic lesion with speckled calcification at the proximal tibia the clinical diagnosis of which favored chondrosarcoma. Incisional biopsy revealed an innumerable number of small globular shapes, whitish parasites. Histologically, the parasites were composed of a few layers of smooth muscle and several calcerous bodies that were enclosed within a single row of tegumental cells. The latter exhibited a wavy appearance and coated with microvilli. These morphologic findings confirmed the nature of these maldifferentiated larvae. The patient was treated by partial resection of the lesion. This should remind clinicians that parasitic infection of the bone can produce a tumor-like lesion.

Key word : Bone, Parasite, Pathology, Intraosseous, Proliferative, Sparganosis

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Sparganosis is an infection caused by a sparganum: the generic term for the second stage larva or plerocercoid of a tapeworm in order Pseudophyllidea[1]. The typical spargana are white, flat, and ribbon-like worms. The size varies from a few millimeters to several centimeters in length[1]. Most cases of sparganosis are found in Oriental countries and are probably caused by species *Spirometra erinacei*, a parasite of carnivors[2]. In North America, most spargana are probably *S. mansonides*, a parasite of cats.

There are three mechanisms of human infection[1]. The first, the most common pathway, is accidental ingestion of procercoids by contaminated drinking water that contains the first intermediate host such as cyclops. After that, the parasites can migrate from the gut and develop into spargana in varying organs. The second pathway is the result of ingestion of a sparganum by eating the flesh of an infected second intermediate host such as fish, frog, or snake. The third is infection by direct contact of an infected second intermediate host to an open wound, allowing the sparganum to crawl directly into human tissue.

The usual sites of infection are subcutaneous tissue[3], subconjunctiva[4,5], and brain[6]. The other sites of human sparganosis include the spinal cord[7,8], intestinal wall[9], breast[10], and scrotum[11].

Rarely, a sparganum will be proliferative, spliting longitudinally and budding profusely, called "*Sparganum proliferum*"[2]. Such a lesion is a very serious condition since they may contain thousands of parasites. The infected organs will conform a honeycomb mass. Less than twenty cases of proliferative sparganosis in humans have been mentioned. To the best of our knowledge, only two cases with skeletal involvement have been reported[12,13], both involved the axial bone. Our present case is the first case of intraosseous proliferative sparganosis that involved the long bone for which clinical and radiographic presentations mimicked a primary bone tumor.

CASE REPORT

A 51-year-old Thai man visited Maharaj Nakorn Chiang Mai Hospital, Chiang Mai, Thailand, complaining of a gradually enlarged mass on his right leg for two years. He had continuous pain on this mass even while resting. No previous history of trauma to his leg was encountered. He had a habit of eating raw meat and often drank fresh water from a well near his house.

There was no history or signs of any systemic illness. The physical examination of the right leg revealed a hard non-tender mass, about 10x5x4 cm, fixed to the anterior part of the tibia, just below the knee joint. The roentgenographic study showed the moth-eaten destruction with spotty calcification thorough the proximal part of the right tibia and also in the expanded mass anteriorly (Fig. 1). The differential diagnoses included chondrosarcoma, Ewing's sarcoma and osteosarcoma. The other laboratory examinations indicated hemoglobin 14 g/dl, hematocrit 42 per cent, leukocyte count 5,600 cells per mm^3: 49 per cent neutrophils, 35 per cent lymphocytes, 11 per cent monocytes, 3 per cent eosinophils and 2 per cent basophils. The blood chemistry included 4.6 mg/dl of serum calcium, 3.1 mg/dl of phosphate, 13.28 U/L of acid phosphatase and 7.6 U/L of alkaline phosphatase. The urinalysis was normal.

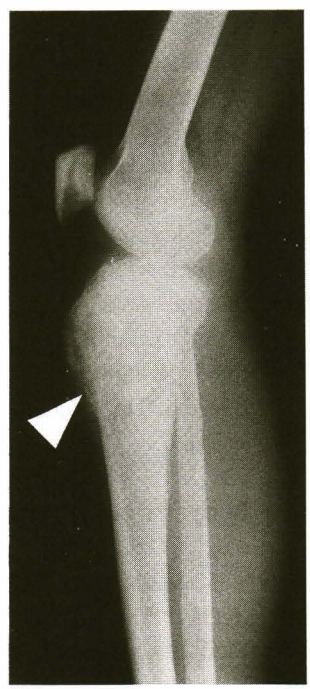

Fig. 1. The plain film right leg, lateral view, shows a bone mass (arrowhead) involving the epiphysial and metaphysial part of the proximal tibia. It destroys the bone in a moth-eaten pattern and breaks through the bony cortex. Focal spotty calcifications are noted.

The incisional biopsies were performed twice followed by marginal excision of the proximal tibia (Fig. 2) and reconstructed by Rush pins incorporated with bone cement.

Pathologic findings

The specimens of the first and second biopsies consisted of few small pieces of gritty, gray white, rubbery to firm tissue. The specimens from marginal excision of the mass consisted of a 14x12x 2.5 cm aggregate of gray white spongy bone and soft tissue from the right proximal tibia, four small pieces of brown muscle, and four small pieces of gray tan soft tissue from the medullary canal of the right tibia. Innumerable small, gray white, oval, globular or multinodular shaped parasites, measuring 0.1 up to 0.8 cm in the greatest dimension, were identified in the specimens. The bone and soft tissue were penetrated by these larvae, being the spongy or honeycomb mass.

Hematoxylin and eosin stained sections from paraffin blocks revealed several parasites in cavities within the fibrous granulation tissue. Some of them were encircled by chronic granulomatous inflammation (Fig. 3A). In the surrounding tissue, there were several lymphocytes, plasma cells, histiocytes and foreign body giant cells with a few eosinophils. Aggregation of foamy macrophages were also noted. The bone marrow was also filled with granulation tissue and the parasites. A few parasites were closely attached to the bony trabeculae (Fig. 3B). The parasites had round, ovoid, tubular or branch shaped in section. They were covered by

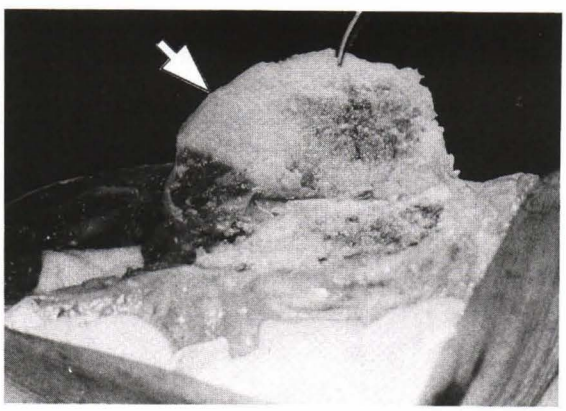

Fig. 2. The operative field demonstrates the mass at the proximal tibia and its cut surface (arrow).

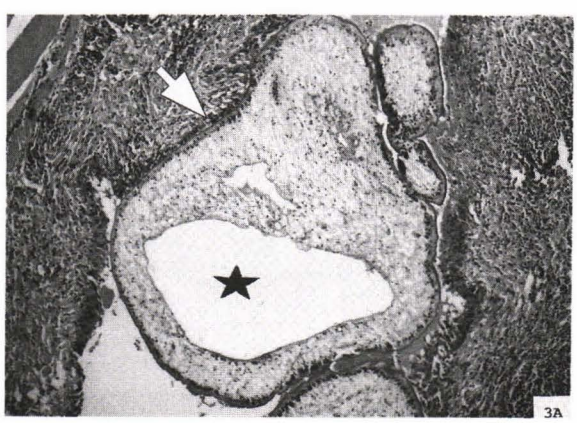

Fig. 3A. Histologic section reveals the cross section of a parasite (white arrow) surrounded by chronic granulomatous inflammation. There are variable sized vesicles (black star) and poorly differentiated parenchymatous tissue. (Hematoxylin and Eosin stain, X40)

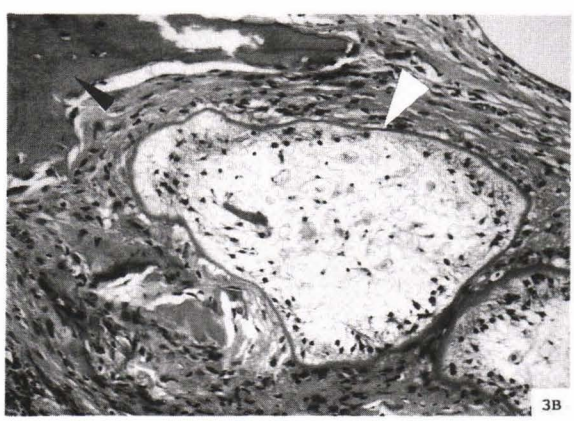

Fig. 3B. The picture shows a bony trabecula (black arrowhead) and the nearby parasite (white arrowhead). (Hematoxylin and Eosin stain, X100)

wavy and eosinophilic tegument, measuring 5 up to 10 micron in thickness (Fig. 3C). Periodic acid-Schiff (PAS) stain showed strongly positive staining along their tegument. Occasionally, the tegument was invaginated to form small cavities called "excretory channels" which were lined by thin membrane (Fig. 3C). Those channels sometimes contained pinkish materials. A single layer of epidermal cells was seen below the tegument. Subtegumental muscle fibers were sparsely distributed in all of the larvae but they were not well developed. Several calcareus bodies were found in the parasitic stroma (Fig. 3D).

Scanning electron microscopic study of the parasites showed globular and nultinodular shaped plerocercoid (Fig. 4A). Their external surfaces were finely granular and partly covered by thin membrane. Irregular infolding of the tegument was generally observed. Numerous round materials were excreted from the excretory pits (Fig. 4B).

DISCUSSION

The first case of human proliferative sparganosis was described in Japan by Ijima in 1904[14]. Since then, less than 20 cases have been reported (13-23) (Table 1). All the patients were adults with a history of drinking fresh water from a natural source or eating uncooked flesh. Most of them were from South East Asia, including those in Japan and Taiwan. The ages ranged from 24 to 62 years with the male to female ratio of 4:3. Their occupations were variable. All of them were sporadic cases.

The most common sites of this parasitic infiltration were skin and subcutaneous tissues. Other less common sites in order included: visceral organs (such as the abdominal cavity[21], pleural cavity[20], and brain[16,18]), bone[12,13], and spinal cord[23]. Skin involvement was usually multiple or disseminated. Sometimes, they produced a large tumor mass, especially in the subcutaneous tissue or muscle.

Parasitic invasion to bone has been found in two cases reported by Liao[12] and Nakamura [13]. The sites of the involved bones were lumbar spine and iliac bone in which the parasites usually destroyed the bony cortex and medullary cavity. Unlike the previous two cases, our case had metaphysis of long bone involvement with speckled calcification. This lesion caused radiologic findings similar to those of chondrosarcoma by showing aggressive destruction of moth-eaten appearance combined with many spotted calcifications. The tiny

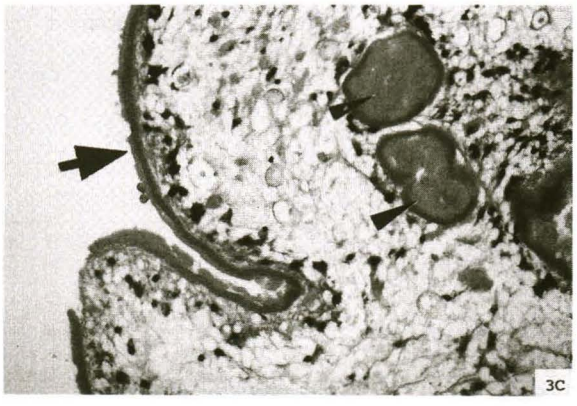

Fig. 3C. Higher magnification of the parasitic wall reveals the linear tegumental layer (arrow) with underneath poorly developed smooth muscle and tegumental cells. Two excretory channels containing pinkish material (arrowhead) are also noted. (Hematoxylin and Eosin stain, X200)

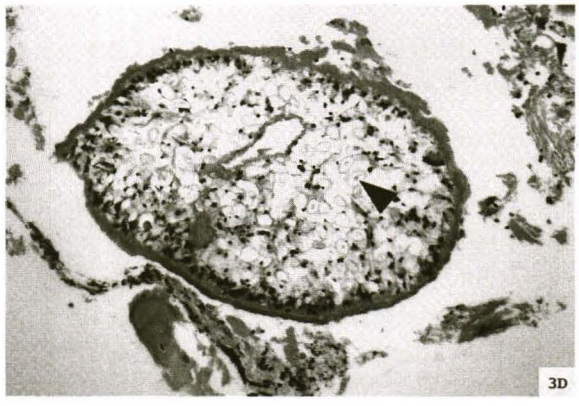

Fig. 3D. Cross section of *Sparganum proliferum* demonstrates the outer most tegument, epidermal cells, sparse muscle fibers, loose stroma, central excretory channel, and several calcareus bodies (arrowhead). (Hematoxylin and Eosin stain, X100)

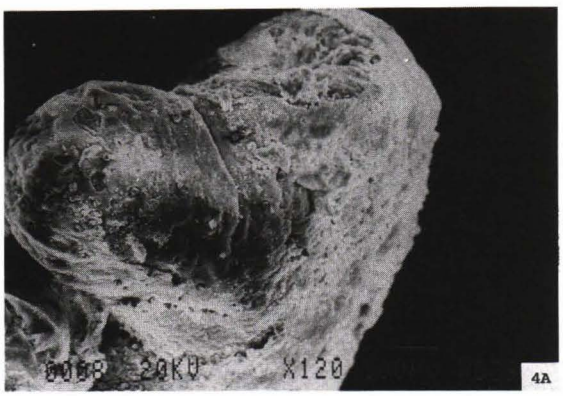

Fig. 4A. Scanning electron microscopic picture reveals a multinodular shaped parasite (X120).

Fig. 4B. Higher magnification of the scanning electron microscope shows the folding tegument and several round shaped materials (arrowhead) at the opening of the excretory channels (x360).

Table 1. Reported 14 cases of human proliferative sparganosis.

Cases			Patient			Site of larva
Author	Country	Year	Age (y)	Sex	Occupation	
Ijima	Japan	1904	33	F	Shopkeeper	Skin of thigh and trunk
Stiles	USA	1908	48	F	Fisherman	Trunk muscles, skin, viscera, brain
Yoshida	Japan	1914	36	M	Monk	Skin, muscles, viscera, brain
Yoshida	Japan	1914	57	M	Unknown	Skin of neck
Morishita	Japan	1920	62	M	None	Skin of chest, viscera
Tashiro	Japan	1924	24	F	Shopkeeper	Skin of neck, viscera, brain
Connor	USA	1976	58	M	Unknown	Disseminated: vessels, lymph nodes, lungs, liver
Lin	Taiwan	1978	28	F	Teacher	Pleural cavity
Beaver	Paraguay	1981	24	M	Farmer, hunter	Abdominal and pleural cavities
Moulinier	Venezuelan	1982	35	M	Unknown	Disseminated: skin and subcutaneous tissue
Liao*	Taiwan	1984	26	F	Teacher	Lumbar spine
Lo	Taiwan	1987	43	F	Housewife	Spinal cord
Makamura*	Japan	1987	47	M	Office worker	Skin of buttock, muscle, pelvic bone
Settakorn*	Thailand	2001	51	M	Farmer	Tibia

* intraosseous proliferative sparganosis

holes in the metaphysis were the penetrated sites of the larvae. The reaction of the bone to this infiltration looked like calcification in calcified tumors. Another extremely rare incidence of parasitic infection reported in bone is echinococcosis[24].

There were two different morphologic features of the larvae. Those reported by Ijima[14] and Stiles[15] were motile vermiform worms with irregular branch, resembling the roots of ginger. The parasite in our cases and four cases in Taiwan[12, 20,23] and Japan[13] were of vesicular or multinodular shape with an underdeveloped muscular system. It is possible that the term *Sparganum proliferum* does not consist of a single species of tapeworm but contains several different larva of the pseudophyllidean family.

Despite the rareness of the disease, proliferative sparganosis has typical macroscopic and microscopic appearances. The parasites can be seen grossly as numerous, small, round, oval, multinodular or ginger shaped gray white semitranslucent structures. Their size varies from 25 microns to 8 mm in the greatest dimension[19,21]. It has the histologic components of a typical sparganum, but the organization and symmetry are lost[1].

The study of the structure and ultrastructure of *Sparganum proliferum* by Noya in 1992[25] exhibited many of the classical tegumental and parenchymal structures previously described for the other larval cestodes. Those structures were either arranged in a distinct fashion or, in some cases, may be completely different. Among the later and of special interest were the single or multiple parenchymal cavities, surrounded by tegument, which in some instances appeared to act as a primitive digestive tract. Their microvilli were also longer than those of the typical spargana[19].

The antigen of *S. proliferum* has not yet been classified, however, the patient's serum reacted with *S. erinaei* antigen by immunoelectrophoresis. Therefore, immunodiagnosis using the antigen of *S. erinacei*[23] can be used as a diagnostic tool for Sparganosis. Medical treatment with antifungal drugs such as Mebendazole and Praziquantel has not been satisfactory[26]. The entire sparganum should be excised. Post-operative radiotherapy is a questionable method of treatment.

In summary, we have demonstrated the first case of intraosseous proliferative sparganosis involving the long bone. This rare parasitic disease can be diagnosed based upon light microscope. The practical significance is that its radiographic findings are similar to those of primary bone tumors especially chondrosarcoma.

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โพรลิเฟอเรทีฟ สปาร์กาโนสิส ภายในเนื้อกระดูก : รายงานผู้ป่วยและทบทวนรายงาน

จงกลณี เศรษฐกร, พ.บ., โอฬาร อาภรณ์ชยานนท์, พ.บ.**, เบญจพร ไชยวรรณ์, พ.บ.*, ปราโมทย์ วนิตย์ธนาคม, วท.ด.*, กำธร ธรรมประเสริฐ, พ.บ.*, สำเริง รางแดง, พ.บ.**

Intraosseous proliferative sparganosis เป็นโรคพยาธิในกระดูกที่พบน้อยมาก ผู้นิพนธ์ได้รายงานโรคนี้ซึ่งพบที่กระดูกชิ้นยาวเป็นครั้งแรก โดยผู้ป่วยเป็นเพศชาย อายุ 51 ปี มีก้อนโตขึ้นช้า ๆ ที่ขาขวา จากภาพถ่ายทางรังสีวิทยาพบรอยโรคแบบ infiltrative osteolytic with speckled calcification ที่ proximal tibia ซึ่งเข้าได้กับโรค chondroblastoma เมื่อนำเนื้อเยื่อของก้อนมาดูด้วยกล้องจุลทรรศน์พบตัวอ่อนของพยาธิจำนวนมาก ผู้ป่วยได้รับการรักษาโดยการตัดก้อนออกบางส่วน แพทย์ผู้ทำการตรวจรักษาผู้ป่วยควรระลึกไว้เสมอว่า intraosseous proloferative sparganosis สามารถทำให้เกิดก้อนในกระดูกที่มีลักษณะเหมือนเนื้องอกปฐมภูมิของกระดูกได้

คำสำคัญ : กระดูก, หนอนพยาธิ, ปาราสิต, พยาธิวิทยา, โพรลิเฟอเรทีฟ, สปาร์กาโนสิส

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