Roentgen Diagnosis of Armillifer Armillatus Infestation (Porocephalosis) in Man*

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ALTHOUGH not uncommon in Central and West Africa, human infestation by the nymphs of Armillifer (syn. porocephalus) armillatus is relatively unknown in the United States.[1] Encysted nymphal forms of Armillifer were found in 7.8% of autopsied patients in the Cameroons and 22.5% in Zaire.[2] Bretland[3] in a review of the world literature in 1962 found 14 cases of calcified nymphs of Armillifer diagnosed radiologically. This total included one patient from the United States, a Phillipino male whose disease was caused by Armillifer moniliformis, a species encountered in Asia.[4] Bretland states that the radiological appearance of calcified nymphs of Armillifer are among the most characteristic of soft tissue calcifications, but because this appearance is generally not appreciated by physicians practicing outside of endemic areas the diagnosis frequently is missed when patients travel to areas distant from their native lands.

Recently, we encountered a patient in whom the typical roentgenographic findings of Armillifer disease were recognized. Because this man was a resident of a large metropolitan city in the Eastern United States, it is not unreasonable to assume that other patients with this disorder currently reside in this country, and that they may, for reasons not necessarily related to their parasitic infestation, present for radiological examination. Accordingly, this case is being presented in the hope of familiarizing radiologists with the roentgenographic manifestations and the possible clinical importance of this disease.

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CASE REPORT

A 35 year old Liberian male was admitted to the Episcopal Hospital of Philadelphia in urinary retention. Aside from symptoms attributable to his lower urinary tract, his past medical history was not significant and review of systems and family history were also unremarkable.

The patient was well developed and well nourished with a distended bladder palpable three inches above the symphysis pubis. Urinary retention was relieved following the dilatation of a urethral structure. The white blood cell count was 6,200 with a normal differential; hemoglobin 13.3 gms and hematocrit 41%. Blood chemistry values, as determined by SMA-12, were normal. A scout film of the abdomen, prior to intravenous urography, revealed numerous crescentic calcifications scattered throughout the abdomen, more numerous in the right upper quadrant (Figs. 1 & 2). Supplemental views showed these to be related to the peritoneal cavity. These calcifications had a characteristic appearance of calcified nymphs of Armillifer. There were no symptoms related to the Armillifer. Aside from the soft tissue calcifications, the intravenous urogram was normal. Cystoscopy revealed multiple urethral strictures and a vesical neck contracture. Transurethral incision of the vesical neck contracture and internal urethrotomy was performed and the patient had an uneventful convalescence.

DISCUSSION

Armillifer armillatus belongs to the class Pentosomida, family Linguatulidae ("tongue-worms"), a group of degenerate

arthropods which are blood sucking endoparasites of vertebrates.[5]

Actually, according to Manson-Bahr, the linguatulids, (pentasomes or "tongue-worms") are neither protozoa nor helminths but occupy an intermediate position between the annelids (worms) and the arthropods. Pentastomids may be elongated, flattened or cylindrical, and they contain external rings that do not correspond to internal segmentation. Adults have no external appendages other then two pairs of hollow retractable hooks near the mouth. Separation of the body into definite regions as in insects does not occur. In fact, there is little resemblance to arthropods except in the mite-like form of the larva.

Fig. 1. Scout film of abdomen showing multiple calcifications.

In a classic article on the Linguatulidae, Sambon[6] listed 13 genera and 43 species of which the following five species were known to be possible parasites of man: Porocephalus crotali, Porocephalus subulifer, Armillifer armillatus, Armillifer moniliformus and Linguatula serrata. Armillifer armillatus and Armillifer moniliformus are usually responsible for the condition known as porocephalosis in humans.

The adult Armillifer armillatus is found in the trachea and bronchi of pythons, cobra and other snakes, in Central Africa and West Africa, especially the Congo and Nigeria. The adult is vermiform, yellowish and translucent. The male is 3-5 cm. long and female 9-12 cm. Armillifer moniliformis is more slender and has more external rings than Armillifer armillatus. The female is oviparous. The eggs, double-shelled and resistant to water and gastric juice, are laid in the posterior portions of the nasal passages of the snake and are released from the body in nasal secretions and in excreta. The eggs embryonate upon reaching damp vegetation or water and are then ingested with contaminated water or food by rats, monkeys, and certain other mammals. Man acquires the infestation by drinking pond water contaminated by snakes or by eating snake meat. The eggs hatch upon reaching the digestive tracts of the various intermediate hosts. The young larvae are liberated by digestive juice action, penetrate the intestinal wall, and come to lie under the peritoneum where they

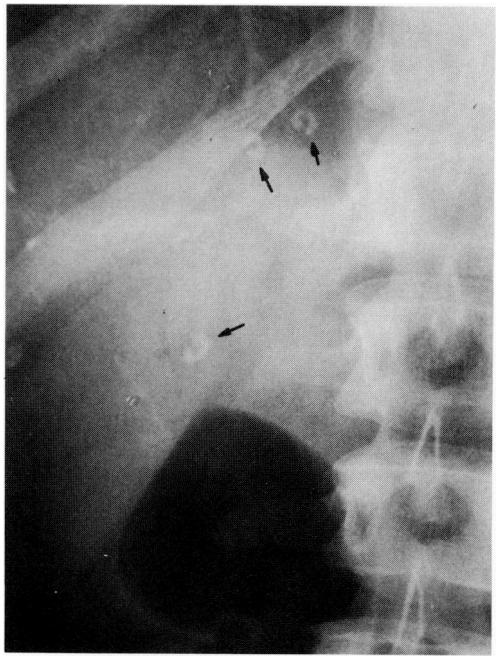

Fig. 2. Unique crescent shape of calcified Armillifer nymphs in right upper quadrant.

undergo several moults to become nymphs. For the life cycle to be completed, it is necessary for the snake to eat the animal containing the ensyted nymphs. The nymphs are liberated in the intestine of the snake; from here, by boring through the mucosa, they enter the mesenteric venules and migrate to the lungs where they become adults.

In man, the cycle comes to an end and the larvae (nymphs) encyst in the liver, intestinal mucosa, lungs and peritoneal surfaces especially over the liver. The nymph lies coiled within the cyst with its ventral surface corresponding to the convexity of the curve. This position is responsible for the characteristic radiographic appearance (Figs. 1 & 2). Pathological changes occur around the parasite, which vary from minimal reaction around live parasites to necrotic granulomatous reaction followed by fibrosis and calcification around dead parasites. When the nymphs are seen "en face", they are said to be like no other pathological or normal calcification, radiologically presenting as horse-shoe or crescent-shaped bodies 4-7 mm in size.[5,7] The lesions are usually multiple but may be single.[8] Differentiation from a calcified cysticercus might present difficulty in the case of a single lesion seen on end; in such circumstances a view in another plane would probably serve to distinguish them.[3] Cysticerci usually occur in skeletal muscle and brain. Bretland reported one case[3] where Armillifer was probably present in the sacrospinalis muscle. Linder found 14 cases of Armillifer nymph calcification in a retrospective analysis of 1,000 abdominal films from Nigerian patients.[9] The most common location was in the right upper quadrant of the abdomen. In Steinbach's case[4], the calcified nymphs of Armillifer moniliformis were present in the thorax, abdomen and scrotum, and were identical in appearance to the nymphs of Armillifer armillatus. Steinbach states that the roentgen appearance of all Linguatulidae is similar regardless of species. The life cycle of Armillifer moniliformis is similar to that of Armillifer armillatus, although the distribution of human disease attributable to this species is limited essentially to the Orient.[5]

Most infestations produce few or no symptoms but in a heavy infestation the migration of nymphs may cause sufficient peritoneal irritation and pain to mimic almost any acute abdominal condition and result in a laparotomy before the true diagnosis is recognized.[10] A blood eosinophilia may occur especially after death of larvae. Severe Armillifer infestation has caused intestinal obstruction, pneumonitis, meningitis, pericarditis, nephritis, peritonitis, obstructive jaundice and even death.[2,5]

Sporadic reports of human disease produced by other linguatulids have been reported. Among these are the wide-ranging Linguatula serrata, which produce Halzoun (Marrara syndrome; parasitic pharyngitis), a disease encountered in Europe and South America; and Porocephalus crotali found in new world rattlesnakes, and responsible, rarely, for human disease.

With increasing world travel Armillifer nymphs will probably be seen more frequently in the United States. It is important that these calcifications be correctly identified, and that symptoms, if present be correctly interpreted in the light of the known clinical and radiologic features of Armillifer disease (porocephalosis).

SUMMARY

A case of Armillifer armillatus infestation (porocephalosis), in a Liberian resident of the United States, is presented. The characteristic radiologic appearance of Armillifer is described, and clinical findings which may be caused by this parasite are reviewed. A brief review of the epidemiology of Porocephalosis is also included.

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