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CASE REPORT

The lord of the broken rings: African porocephaliasis, an emerging radiological diagnosis in developed countries

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African porocephaliasis is a pathology encountered among natives of tropical African countries. While clinical symptoms caused by parasite migration are usually encountered in tropical countries [1, 2] clinicians and radiologists can be confronted with calcified larval parasites detected on X-rays performed for unrelated symptoms [3, 4]. We hereby report two cases of porocephaliasis discovered fortuitously on abdominal and chest X-ray in two African patients seen in medical consultation.

Case 1

A 16-year-old Cameroonian girl was admitted for multiple calcifications fortuitously discovered by means of thoracic abdominal X-ray performed on her arrival in France (Fig. 1). Her medical history was uneventful, except for malaria and typhoid fever. Clinical examination and standard biological tests were normal. Parasitological serology tests were negative for hydatidosis, bilharziasis, toxocariasis, cysticercosis, and distomatosis; and were equivocal for strongyloidiasis, filariasis, and trichinellosis. Parasitological examination of stool samples was negative, and no mycobacteriae were found in the sputum or urine The calcifications' thoracoabdominal distribution, especially involving the liver, and their broken-ring shape were highly suggestive of African porocephaliasis. The patient denied snake consumption.

Case 2

A 40-year-old male native of the Central African Republic underwent X-ray for non-specific symptoms consisting of chest pains due to a recent physical aggression in Africa. A small number of broken-ring shaped calcifications were revealed, exclusively located in the abdomen (Fig. 2). As in Case 1, standard biology parasitological serology and stool analysis were normal. The porocephaliasis diagnosis was based solely on the corresponding characteristic calcifications observed on X-ray. It should be noted, however, that in this case the patient reported having consumed snake at home.

Pentastomida are annulated, non-segmented vermiform blood-sucking endoparasites that share features with both *Arthropoda* and *Annelida*, though they are typically known to form their own phylum. *Pentastomida* comprise two orders: *Porocephalida* and *Cephalobaenida*. *Porocephalida* includes two families of medical interest: *Porocephalidae* and *Linguatulidae* (tongue worms). Six species are known to infect humans: *Armillifer armillatus, Linguatula serrata, Armillifer moniliformis, Armillifer grandis, Leiperia cincinnalis,* and *Raillietiella hemidactyli. A. armillatus* and *L. serrata* account for over 99 % of all reported Pentastomiasis cases [4]. The human diseases related to these parasites are known as pentastomiasis, further categorized as porocephaliasis for *Armillifer* infections and linguatuliasis for *Linguatula* infections. Porocephaliasis is a tropical infection caused by two African species, namely *Armillifer armillatus* and *Armillifer grandis*, and one Asian species: *Armillifer moniliformis*. A new species, *Porocephalus taiwana,* was described in south-east Asia in 2005 in the watery stool of a patient [5]. The exact incidence of African porocephaliasis

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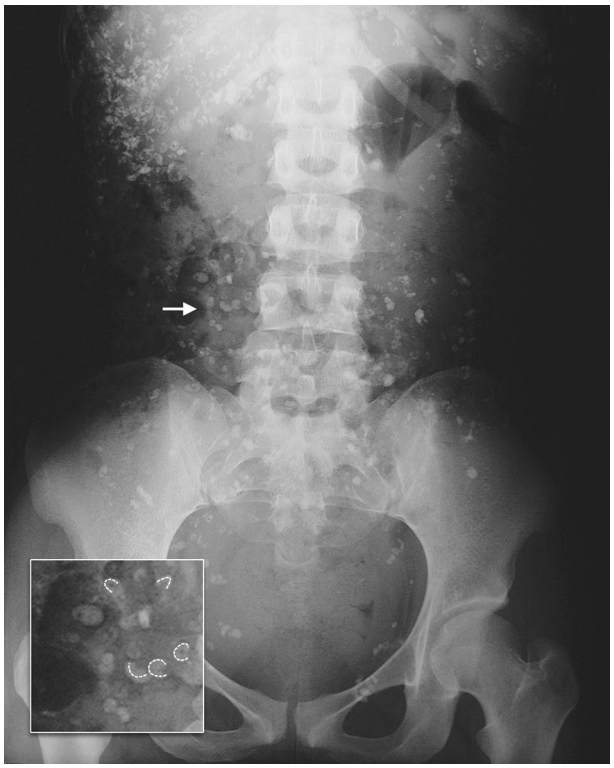

Fig. 1 Case 1. Multiple disseminated abdominal *broken-ring* shaped calcifications, (*arrow*) indicative of porocephaliasis, in a 16-year-old female native of Cameroon

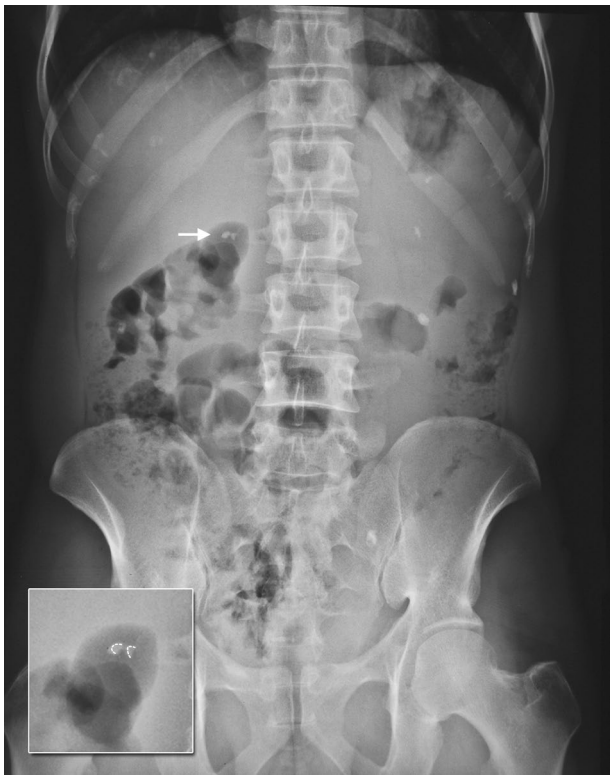

Fig. 2 Case 2. Abdominal calcifications (*arrow*) indicative of African porocephaliasis in a 40-year-old male native of the Central African Republic

is not precisely known, yet appears common, with prevalence ranging from 1.4 % in Nigeria and 2.1 % in Cameroon to up to 26.6 % in the Democratic Republic of Congo [6, 7]. A higher prevalence has been described among members of the same family or within one ethnic group that consumes snakes [6]. The adult parasites live in the respiratory tract of constrictor snake species, such as pythons or bitis vipers. Humans, representing a dead-end for the parasite, are usually accidently infected by swallowing eggs from food contaminated by the dejections or sputum of these snakes [6], or by eating (or handling) uncooked snakes, which can lead to massive infestation [4]. The eggs hatch and liberate larva, which migrate before settling primarily in the organs of the peritoneum, mesentery, liver, lungs, or lymph nodes, then transforming into encysted nymphs. Humans typically suffer little discomfort from this *larva migrans*, with the encysted nymphs becoming calcified in less than 2 years. While transitory eosinophilia is at times noticed, it nevertheless remains either unexplained or related to another parasite [3]. Several clinical symptoms related to motile larva have, however, been observed: peritoneum irritation and occlusion or lesions in organs such as the eyes or brain, leading to medical emergencies and even death in rare cases [2, 4, 6, 8–10]. Parasites burden may greatly differ between patients as illustrated by our two cases. Massive infestation have been described in case of ingestion of a parasite female full of eggs with potentially fatal issue [2]. Patients may; however, harbor hundreds of larvae without symptom and host-parasite interactions deserve future research attention [11]. The calcified parasites are typically asymptomatic [12], though abdominal pain has been reported [13]. The diagnosis and treatment of "young" non-calcified stages of porocephaliasis are difficult, since its clinical and biological manifestations are unspecific at this stage, and the only reliable option involves surgery to excise and test for the parasites. Surgical procedures resect nodular structures of 1–2 cm for histological examination, which reveals encapsulated *Armillifer* nymphs [1, 14]. Serology tests have previously been developed but none are commercially available and they are only performed in reference laboratories with homemade assays [15]. Most African cases are, however, discovered incidentally during X-ray investigations [9]. In Europe and North America, cases are rare and most often accidentally discovered at a calcified stage by means of X-ray, computed tomography (CT) scan, surgery or autopsy, always involving African migrants who are examined for unrelated symptoms [3, 4, 16]. On X-ray, the characteristics of porocephaliasis are notable for multiple calcifications, sometimes hundreds, which are predominantly localized in

the hepatic area [3, 4]. The shape of porocephaliasis calcifications is also quite characteristic and is the cornerstone of the diagnosis at this stage: they measure approximately 1 cm and are curved like broken rings (also described as a C-shaped horse shoe), though other shapes have been described, such as a comma, dense circle or ring. These calcifications differ from those of other parasites such as those involved in cysticercosis or trichinosis infection which are localized in the muscles and measure approximately 1 cm in length. The principal differential diagnosis is cysticercosis a parasitic infection caused by the cestode (tapeworm) *Taenia solium* following the ingestion of undercooked pork. Most patients with cysticercosis have muscle and subcutaneous localizations without symptoms and are diagnosed incidentally on X-ray when cysticerci are calcified. However, the localization and the shape of calcifications are different from porocephalosis with presence of multiple "dots", "rice-grain" or "cigar-shaped" calcifications in the muscle bundles [17]. Echography and CT scan can reveal the precise location of porocephaliasis nymphs [13].

The increasing numbers of African migrants living in developed countries and their access to the health system of these countries, especially standard X-ray examinations [18] will result in an increase in diagnosis of asymptomatic porocephaliasis at a calcified stage, as observed in our unit. Clinicians should be aware of this neglected tropical disease to avoid unnecessary and costly examinations.

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Compliance with ethical standards

Conflict of interests The authors declare that they have no conflict of interest.

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