

REVIEW

Imported *Armillifer* pentastomiasis: Report of a symptomatic infection in The Netherlands and mini-review



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Summary We report a case of symptomatic visceral *Armillifer* pentastomiasis in a 23-year-old female Liberian immigrant to The Netherlands. The patient was referred to the gynecologist because of lower abdominal pain. During laparotomy, multiple adhesions were seen in the lower pelvis and a hydrosalpinx with an encapsulated *Armillifer* nymph, most likely *Armillifer armillatus*, was found. Key features of the parasite's cuticle which facilitate the diagnosis of pentastomiasis, are presented. Symptomatic pentastomiasis is uncommon, and most cases are diagnosed incidentally during surgery for other reasons, or at autopsy. With regard to increasing international migration, other imported pentastomiasis cases to Europe and North America are reviewed, and more cases are likely to be seen in the future.

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Introduction

Visceral pentastomiasis is a zoonotic parasitosis with an increasing number of documented human infections by the larval stages (nymphs) of pentastomes (tongue worms) [1,2]. The vermiform organisms are related to branchiuran crustaceans [2,3] and possibly co-evolved with their respective final hosts [2]. Adult pentastomes inhabit the respiratory tract of their final hosts, which are large tropical snakes in the case of *Armillifer* sp., and infective ova are discharged into the environment with host respiratory secretions and feces. Mammals, such as rodents, small monkeys, and accidentally humans, become intermediate hosts when they ingest the parasite ova with contaminated food or water. In the intestine of the intermediate host, the larvae hatch, migrate to the viscera, encyst and mold several times [1]. The life-cycle of *Armillifer* sp. is completed when the natural intermediate host with the encapsulated pentastome larvae is eventually eaten by the final host (Fig. 1). In humans, *Armillifer* larvae are found in the serosa around abdominal organs, abdominal lymph nodes, in the liver parenchyma, the mesenterium and intestinal wall. Sometimes the pleura or the lung parenchyma is infected [1]. Despite an occasional large amount of nymphs found in human viscera, the infections are usually asymptomatic and pentastomiasis is an incidental finding during surgery or at autopsy [1]. Besides contaminated food or water, risk factors for human *Armillifer* infection include the handling and consumption of contaminated snake products, including meat and bile [1,2]. Tropical snake farming may also become a public health concern [2].

Most human infections are caused by *Armillifer armillatus*, a parasite found in West and Central Africa, and only rarely by *Armillifer grandis* in Central Africa [1,4]. The majority of African cases are described from Nigeria and the Congo region [4]. Pentastomiasis is also documented in Ghana, reflecting the socio-cultural habit of totemism, with the python being central to this practice for many tribes in the savanna belt [5]. In China, *Armillifer agkistrodontis* is prevalent [6], whereas *Armillifer moniliformis* occurs in South-East Asia [1,7]. The latter species has recently re-emerged causing a human infection after nearly 40 years in Malaysia [7]. Only rarely, infections in immigrants to Europe and North America are noted (Table 1). Imported African pentastomiasis to The Netherlands has not been reported so far.

Here, we describe a case of symptomatic pentastomiasis in a female patient from Liberia, West Africa, who developed abdominal pain due to a nymph of *Armillifer* after the arrival in The Netherlands. In the light of increasing international migration, other imported cases to Europe and North America are reviewed.

Case report

In 2005, a 23-year-old woman from Liberia was referred to the outpatient clinic of the gynecology department of a peripheral hospital in The Netherlands. The patient complained about severe abdominal pain in the left lower quadrant. She had immigrated to The Netherlands 16 days before, and, after arrival, she had requested asylum because of repeated sexual abuse. The pain had existed in



Figure 1 Life-cycle of *Armillifer* spp. Large snakes, such as pythons, are the final hosts for adult *Armillifer* pentastomes, harboring the parasites in their airways. With the snakes' secretions and feces, infective parasite ova are shed into the environment, and accidentally taken up by mammalian intermediate hosts (rodents and monkeys). In these intermediate hosts, the larvae hatch from the egg and encyst in the viscera. There, the parasite larvae wait for the intermediate host to become prey of snakes, and the developmental cycle is completed.

Table 1 Imported cases of tropical pentastome infections to Europe and North America.

Year	Patient's origin	Immigrated to/place of diagnosis	Method of diagnosis	Organ systems involved; symptoms	Reference
1976	Liberia	United States	X-ray	Abdomen; urinary retention (not related to pentastomiasis)	Mapp et al. [29]
1982	Africa	France	X-ray	Abdomen; no symptoms	Piéron et al. [30]
1991	Nigeria	Canada	Autopsy	Pleura, lungs, liver, mesentery, peritoneum; no symptoms reported	Guardia et al. [31]
1999	Congo	France	Autopsy	Lungs, liver, mesentery, intestine; Paresis, daysarthria, seizures (attributed to hypersensitivity as no parasites were found in the brain)	Lavarde et al. [12]
2004	Mali	France	X-ray	Liver, spleen, peritoneum, small bowel and colonic walls; No symptoms	Sellier et al. [32]
2005	Nigeria	Spain	Laparoscopy	Retroperitoneum close to the uteroscral ligament; abdominal pain	Martín-Rabadán et al. [11]
2013	Togo	Germany	Autopsy	Liver; no symptoms	Tappe et al. [4]
Present report	Liberia	The Netherlands	Laparotomy	Abdomen; Hydrosalpinx	Tappe et al.

Except for the X-ray diagnoses, by which the several *Armillifer* species cannot be discriminated from each others [33], the species involved was *A. armillatus*. Other snake-associated pentastomes, such as New World *Porocephalus* sp., have only rarely been responsible for human disease [29], if ever [34]. The latter name has often been used synonymously for many pentastome species. *Linguatula serrata* is a cosmopolitan canine-associated parasite, and has thus not been included in the table though imported infections exist [35].

a lower intensity for a longer time period already, and had recently exacerbated. The last menstruation had occurred two weeks before. Detailed medical history could not be obtained due to language barrier. On examination, diffuse pain was felt in the lower abdomen. The right adnex had no palpable abnormalities, while the left adnex could not be examined manually due to active defense. Colposcopy showed a normal portio, and transvaginal echography disclosed a normal uterus with an unremarkable endometrial thickness. Left behind the uterus, a cystic echogenic structure of 3.9 × 4.2 cm was visible without Doppler flow signals. Based on these findings and the acute pain, a diagnostic laparoscopy was intended. However, during the procedure, the internal genital organs showed multiple adhesions, and an open laparotomy was performed instead. After bilateral adhesiolysis a left-sided hydrosalpinx, which was completely attached to the uterus, and a cystic encapsulated C-shaped structure of possible helminth origin was found. The hydrosalpinx was opened and a fresh hemorrhage was discovered. The cyst was resected for parasitological examination. The patient recovered rapidly without medical problems and the pain subsided.

Visual examination of the encapsulated parasite revealed a C-shaped vermiform body of approximately 8 × 3 mm in diameter. The outside of the cyst was rough with a deposit of brownish round calcified structures (Fig. 2A). Microscopic examination of the parasite body showed pores in combination with long calcified stretched cells on the outside (Fig. 2B and C), characteristic for the sclerotized openings in the cuticle of pentastome nymphs [1]. The morphological features of the parasite and the origin of the patient were consistent with the diagnosis of visceral pentastomiasis due to a nymph of *Armillifer* sp., most likely *A. armillatus*. However, the inner structures of

the parasite were completely degenerated and the number of annulations could not be determined. A polymerase chain reaction investigation targeting the 18S rRNA gene of pentastomes [2,4,8] performed on the formalin-fixed and calcified remains of the parasite was negative. No serum was stored for retrospect pentastomiasis serology. The patient reported frequent close snake sightings in her home country, but direct snake contacts were denied.

Discussion

Most human cases of pentastomiasis are asymptomatic and incidental findings [1]. The clinical manifestations of pentastomiasis have a great variety and depend on the tissues and organs infected. Living, intact and encysted pentastome nymphs produce only little or no symptoms, whereas dying, antigen-releasing parasites provoke clinical illness [1]. Blood eosinophilia may be present [1], especially after death of the larvae. Clinical manifestations of visceral pentastomiasis include abdominal pain, chronic cough, or night sweats [1]. Abdominal emergencies, as seen in our case, have been described [9–11]. In severe infections with *Armillifer* species, death may occur due to disseminated disease/hypersensitivity [12,13], secondary septicemia [14], severe enterocolitis [15] and intestinal obstruction [16]. In our case, abdominal pain was caused by a hydrosalpinx which was in turn possibly due to two factors: a pre-existing parasite pathology close to the left adnex which was presumptively aggravated by the repeated sexual abuse.

Diagnosis of visceral pentastomiasis can be achieved by radiology, showing calcified nymphs with a horse shoe or C-shaped structure on pulmonal or abdominal radiographs [1].

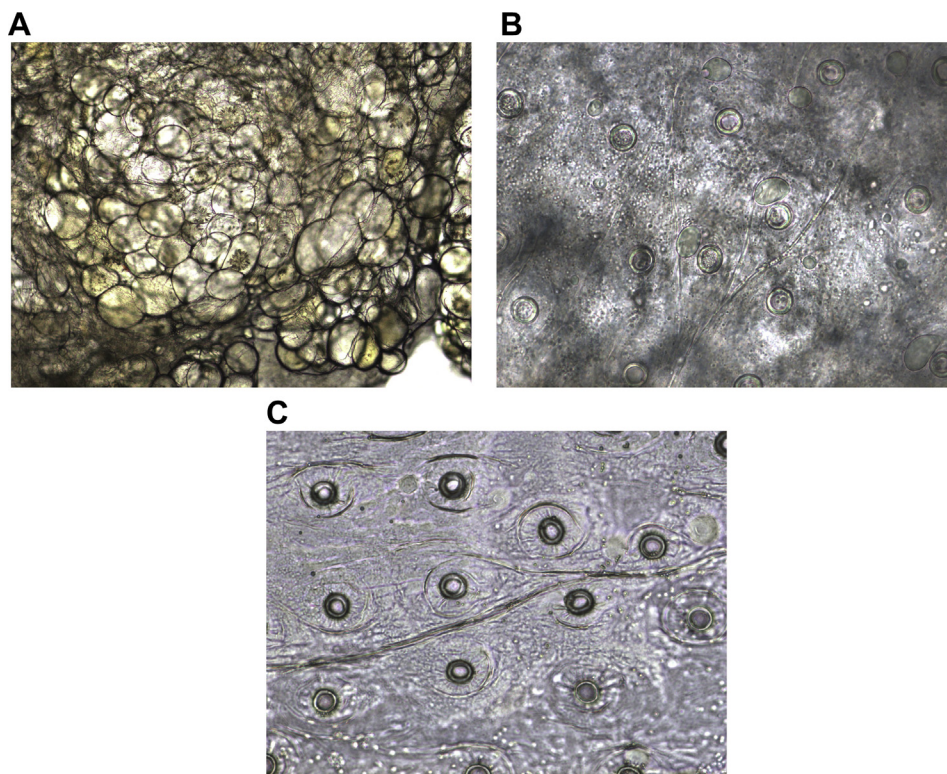


Figure 2 High magnification of the encysted pentastome nymph and the parasite's cuticle. **A**, View on the outside of the parasite's cyst. A multitude of round brownish particles are visible which are possibly calcium deposits. Pentastome nymphs molt several times within their fibrotic cyst. The shed cuticle (exuvia) remains in close contact to the parasite, calcifies and is decay-refractory. Original magnification $\times 100$. **B** and **C**, Detail of the parasite's chitinous cuticle. The cuticle is penetrated by ring-like structures, which are openings of the ducts of subcuticular glands [1]. The openings are sclerotized (best seen in **B**) and surrounded by a radially striated structure (**C**) which is embedded in long stretched cells. Original magnification $\times 400$. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

Serological assays for human infection have also been developed [2,17], but no serum was stored for retrospect analysis in our case. Recently, PCR has been successfully used for species diagnosis from a formalin-fixed, paraffin-embedded tissue sample in a human case [4], as well as in several veterinary cases [2,8,18]. In the case presented here however, the specimen was too degenerated to allow a positive PCR result. Otherwise, diagnosis of visceral pentastomiasis is based on gross pathology or histology. In our case the cuticular structures of the resected entirely calcified parasite, which can easily be seen under a stereomicroscope, facilitated the diagnosis. Histologically, three types of pentastome lesions have been described, depending on the degree of degeneration of the parasite [1,19]. Species identification is achieved by counting the annulations (*A. armillatus*: 18–22; *A. grandis*: >25; *A. moniliformis*: approx. 30; *A. agkistrodontis*: 7–9) and measuring the size of the larval parasites (9–23 mm body length, depending on the species) [1,2,20]. In our case, the number of annulations could not be determined, but the size of the parasite, though calcified, matched with *A. armillatus*.

Human visceral pentastomiasis occurs in rural populations with close contact to snakes. Radiological prevalence studies showed rates of 1.4% in Nigeria [21], and less than 1% in the Congo [22]. These rates were considerably

lower than those found in a single serostudy performed in the Ivory Coast, which demonstrated a seroprevalence of 4.2% [17]. However, much higher prevalences were recorded in autopsy series, such as in Cameroon (7.8–12.6% [23,24]), the Congo (12–22.5% [25,26]) and Nigeria (33% [27]). The highest rate, 45.5% in adult Aborigines, was noted in a large autopsy series from Malaysia for *A. moniliformis* [28]. As radiology only detects calcified nymphs and serology possibly only decaying, antigen-releasing parasites, the highest rates from autopsy studies, by which theoretically all nymphs are discovered, are not surprising. With regard to therapy, it is important to realize that in asymptomatic patients treatment is not necessary, since the parasites degenerate after approximately two years [1]. Only in symptomatic infections with numerous parasites a surgical approach may have to be considered. There is no antiparasitic chemotherapy available for pentastomiasis [1]. A few cases of imported visceral pentastomiasis due to *Armillifer* sp. have been described, all except one originated from West Africa [4,11,12,29–32]. Some of them were symptomatic [11,12]. Diagnosis was achieved by radiology, autopsy, or surgery (Table 1). With the increase in international travel and migration, more imported cases are likely to be diagnosed [4]. Potentially, the disease could also be observed in long-term travelers [1].

Conflict of interest

None.

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None.

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