8.13 Pentastomiasis (porocephalosis, linguatulosis

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ESSENTIALS Pentastomiases or porocephaloses are emerging zoonotic infec- tions caused by branchiuran maxillopod crustacean parasites. Linguatula serrata (‘tongueworm’)— cosmopolitan, infecting upper respiratory tracts of the definitive hosts, canids. Nymphs discharged in nasal secretions are taken up by herbivorous animals, the inter- mediate hosts, which pass on the infection when they are eaten. Humans may be infected by eating raw liver and other offal of sheep, goats, and other animals, soon after which acute allergic obstructive naso-laryngo-pharyngitis (halzoun or marrara syndrome) may de- velop. Larvae can be found in sputum and vomitus. Armillifer spp.—these are confined to Africa and Southeast Asia, where they infect the respiratory tracts of snakes. Humans are in- fected by drinking snake-polluted water, eating raw snake, or handling snakes, a common practice in some communities. Most infections are asymptomatic, but massive infections may produce symptoms of an acute abdomen and are rarely fatal by causing in- testinal obstruction or enterocolitis. Nymphs are detected at lapar- otomy or autopsy and (calcified) on abdominal radiographs. Treatment and prevention—aside from standard measures for hypersensitivity phenomena, there is no specific treatment, although praziquantel,

mebendazole, and albendazole have been tried. Surgical intervention may be necessary in cases of obstruction and with intraocular infections. Prevention is by thoroughly cooking all meat of whatever origin, by boiling drinking water and by avoiding intimate contact with snakes.
Introduction Pentastomida are dioecious, obligate parasites that are cur- rently grouped in subclass Branchiura (fish lice and cycloids) of class Maxillopoda, subphylum Crustacea, phylum Arthropoda. Common names are ‘pentastomes’ (referring to two pairs of hooks above the mouth that give the impression of five stomata; Fig. 8.13.1) or ‘tongue worms’ (alluding to the tongue-like appearance of some, such as adult Linguatula). Adult pentastomes inhabit the upper re- spiratory tracts of their end hosts (vertebrates such as reptiles, fish, birds and mammals) where they feed on blood and other tissues. Their larvae infect internal organs of vertebrate or arthropod inter- mediate hosts. Pentastomes appear to have coevolved with other maxillopodan/branchiuran parasites and their vertebrate hosts. There are about 100 living species in the orders Cephalobaenida (e.g. genus Raillietiella) and Porocephalida (e.g. genera Linguatula, Armillifer, Porocephalus, Leiperia, and Sebekia). Humans are ac- cidental dead-end hosts to some nine species of these emerging zoonotic parasites, causing infections termed pentastomiasis, porocephalosis, linguatulosis, or linguatuliasis. Ninety percent of human visceral pentastomiasis is attributable to Linguatula serrata 8.13 Pentastomiasis (porocephalosis, linguatulosis/linguatuliasis,
or tongue worm infection) David A. Warrell Fig. 8.13.1 Adult pentastomid showing mouth (arrowed) and lateral hooks giving the appearance of five stomata. Scanning electron micrograph, ×400. Courtesy of Professor Viqar Zaman.

8.13 Pentastomiasis 1583 or Armillifer armillatus. Nasopharyngeal pentastomiasis (‘Halzoun’ or ‘Marrara syndrome’) is caused by L. serrata. Pentastomiasis used to be regarded as a rare condition, but it is emerging as an increasingly important zoonosis in Africa (e.g. in Nigeria and DR Congo) and China, while imported cases are being reported in Europe and North America in immigrants from Africa. Aetiology Linguatula species Linguatula serrata occurs in Europe, the Middle East, Africa, and North, Central, and South America. The names ‘linguatula’ and ‘tongueworm’ reflect the 72–92 annular grooves and flattened shape, particularly of the adult female. Dogs, foxes, and wolves, the defini- tive (final) hosts, harbour adults and nymphs in their upper respira- tory tract and shed them in their nasal secretions, saliva, and faeces. Rodents and other small herbivorous mammals ingest the ova with vegetation. They hatch in the lumen of the gut, releasing larvae that moult, burrow into the tissues and encyst. When these intermediate hosts are eaten by definitive carnivorous hosts, nymphs hatch from the cysts and migrate to the lungs and nasopharynx where they mature. Clinical features Visceral pentastomiasis When humans ingest ova of Linguatula, such as by eating uncooked meat, larvae hatch in the gut, burrow through its wall, migrate through the tissues, and encyst especially in the liver. Second- or third-stage larvae cause symptoms through obstruction or com- pression (e.g. in biliary, gastrointestinal, or respiratory tracts, men- inges, eye, or brain). Visceral pentastomiasis is often asymptomatic but fever, abdominal pain and distension, ascites, and anorexia have been reported with hepatomegaly, peripheral neutrophil and eosino- phil leucocytosis, raised serum IgE, and anaemia. At laparotomy, autopsy, or by imaging, multiple small sharp-bordered nodules are found in lungs, liver, and abdominal lymph nodes. Nasopharyngeal pentastomiasis (Lebanon ‘halzoun’,
Sudan ‘marrara syndrome’) Ingestion of cysts containing third-stage larvae in raw liver or lymph nodes from sheep, goats, cattle, camels, and lagomorphs can cause nasopharyngeal pentostomiasis, known as ‘halzoun’ (halzun means ‘snail’ in Arabic) in Lebanon and ‘marrara syndrome’ in the Sudan. This has also been reported from Greece, Turkey, North Africa, Egypt,

Jordan, Iran, and elsewhere. In the human stomach, larvae escape from the cysts and migrate up the oesophagus to the naso- pharynx mucosa. Within minutes to a few hours of eating the in- fected viscera, there is intense irritation of the upper respiratory and gastrointestinal tracts causing coughing, sneezing, rhinorrhoea, retching, vomiting, lacrimation, haemoptysis, epistaxis, cervical lymphadenopathy, transient deafness, difficulty in speaking, dys- phagia, wheezing, dyspnoea, and oedema of the face and oro- pharynx. The larvae, which are 5–10 mm long, can be found in sputum and vomitus. Patients usually recover in 1 or 2 weeks, but fatal acute upper airway obstruction is reported. Clinical features suggest a hypersensitivity reaction. Flukes (Fasciola hepatica and Dicrocoelium dendriticum) and nematodes (Mammomonogamus laryngeus) ingested in raw sheep and goat liver, and aquatic leeches (Limnatis nilotica and Dinobdella ferox) (see Chapter 10.4.2) have been implicated in halzoun but cannot explain the classic syndrome. Very rarely, larvae may mature to adulthood in the human nasal cavity, causing bleeding and obstruction. Armillifer and Porocephalus species These are annulated parasites (Fig. 8.13.2a). Adult males and the much larger females (up to 20 cm long) inhabit the respiratory and digestive tracts of snakes (Fig. 8.13.3), especially those of the genera Python, Lamprophis/Boaedon (African house snakes), Naja (cobras) (Fig. 8.13.4), Bitis (African vipers) (Fig. 8.13.2b), Bothrops (Latin American lanceheads) (Fig. 8.13.5), Crotalus (North American Fig. 8.13.3 Whip snake Demansia atra (Papua New Guinea) bringing up a pentastome. Copyright Mark O’Shea. (a) (b) Fig. 8.13.2 Armillifer armillatus. (a) Left: two adults found in the lungs of a rhinoceros viper; Right: calcified nymph from the mesentery of a Ghanaian patient. (b) Rhinoceros viper (Bitis rhinoceros). Copyright DA Warrell.

section 8 Infectious diseases 1584 rattlesnakes) and other vertebrates. Ova are shed in the snake’s nasal secretions and faeces and are picked up by rodents, monkeys, other small herbivorous mammals, and geckoes. Larvae encyst in the tis- sues of these intermediate hosts and will develop to the nymph stage if ingested by another animal, but develop to adults only in snakes. Human infections Humans are infected by drinking water contaminated with ova or by ingesting living encysted larvae in raw or undercooked snake meat, blood, or gallbladders or by handling snakes either in tropical snake farms or in villages in parts of West Africa where they kept as sa- cred totems. Raw snake meat is eaten habitually or as part of ju ju rituals in Africa (Nigeria, Côte d’Ivoire, Benin, Cameroon, and DR Congo) and in South-East Asia, especially by the Temuan tribe of Malaysian aborigines. Ingested ova hatch in the gut, releasing larvae which moult, burrow into the tissues where they encyst as nymphs and usually degenerate after a few years. Epidemiology The prevalence of infection can be judged by discovering calcified nymphs (Fig. 8.13.2) on radiographs of the abdomen and chest (Fig. 8.13.6). These appear as discrete, crescent-shaped, soft tissue calcifications, 4 to 8 mm in size. In West Africa they are seen par- ticularly in the right upper quadrant and are situated beneath the peritoneum covering the liver. In Ibadan, Nigeria, they were seen in 1.4% of randomly selected straight abdominal films (7% in men aged 50–59 years) and in DR Congo in 1%. However, the preva- lence of encysted nymphs or larvae at autopsy was 12–22.5% in DR Congo, 33% in Nigerian patients dying of malignancy, 7.8–12.6% in Cameroon, and 45.5% among Malaysian Orang Asli. Cysts are (a) (b) Fig. 8.13.4 (a) Pentastomes from the lungs of (b) an Egyptian cobra Naja senegalensis. Fig. 8.13.5 Pentastomes found in the respiratory tract of lancehead vipers Bothrops spp., Manaus, Brazil. Copyright DA Warrell. Fig. 8.13.6 Typical radiographic appearance of calcified nymphs of Armillifer armillatus in the abdominal cavity of a Ghanaian patient. Courtesy of Dr GM Ardran.

8.13 Pentastomiasis 1585 found most commonly in liver (Fig. 8.13.7), mesentery, gut wall, peritoneum, spleen, kidneys, omentum, and lungs. In Ibadan, pentastomiasis was the third most common cause of hepatic cirrhosis. In Côte d'Ivoire, seroprevalence was 4.2%. Recently, an epidemic of A. grandis infections was described in Sankuru district of DR Congo. Human infections with the larvae or nymphs of the following species of Armillifer have been reported: • A. agkistrodontis—(length 10 mm, 7–9 annular rings) China (in the snake Deinagkistrodon acutus) • A. armillatus —(length 9–23 mm; 18–22 annular rings) West and Central Africa (Senegal, the Gambia, Ghana, Benin, Nigeria, Cameroon, DR Congo, Zimbabwe), Egypt, and the Arabian Peninsula • A. grandis—(length 9–15 mm, >25 annular rings) Central Africa (DR Congo) • A. moniliformis—(length 12–20 mm, 30 annular rings) Asia (Malaysia, Borneo, Philippines, Indonesia, Tibet, Australia). Intermediate hosts include monkeys, otters, rats, house geckoes, and cockroaches • A. najae—India Human infections with the following species of Porocephalus have been reported: • P. crotali—(38–40 body segments) • P. taiwani—(length 4–5 mm, 10–11 spiral rings) Clinical features Most infections are entirely asymptomatic. However, migration of large numbers of larvae from the gut into the tissue and their deaths, releasing parasite antigens and provoking hypersensitivity, may produce a variety of symptoms including fever, night sweats, persistent cough, abdominal pain and tenderness, vomiting, diar- rhoea, and obstructive jaundice. Some patients have mild blood eosinophilia. Massive infection, perhaps following ingestion of a gravid female, can cause acute abdominal symptoms prompting laparotomy at which hundreds of wriggling nymphs may be dis- covered beneath the visceral peritoneum covering the liver and spleen, in liver parenchyma, mesentery, intestinal wall, abdom- inal lymph nodes, lungs, or pleura. Serious inflammatory and obstructive effects have been described in the gut, peritoneum, mesentery, liver and biliary tract, lungs, pleura, pericardium, myocardium, central nervous system, and eye (see next). These may be due partly to hypersensitivity. The few reported fatal cases resulted from massive liver infections, mechanical intestinal ob- struction, or haemorrhagic enterocolitis complicated by secondary Gram-negative septicaemia. A suggested association between Armillifer infection and colonic or other malignancies has not been substantiated. Ocular pentastomiasis About 20 cases have been reported from the United States, Ecuador, Europe, Israel, India, Brazil and DR Congo (L. serrata, A. armilla- tus, A. grandis). Adnexal infections (e.g. subconjunctival, associ- ated with peri-orbital oedema) were easily removed but intraocular infections usually caused blindness through iritis and uveitis with goniosynechiae and secondary glaucoma, iridodonesis and lens subluxation, and vitreous, subretinal, or retinal detachments. Early surgical extraction of nymphs is recommended. Other pentastomid infections Human infections with Leiperia cincinnalis have been described in Africa. Subcutaneous infections by Raillietiella gehyrae and R. hemi- dactyli occur in Vietnam and by Sebekia species in Costa Rica. In Vietnam, infection with Raillietiella spp. results from swallowing small live lizards for medicinal purposes. Diagnosis The radiographical appearances of calcified pentastomid nymphs are distinctive (Fig. 8.13.6). They are not found in muscle, distinguishing pentastomiasis from cysticercosis. Pentastomes may be discovered at laparoscopy, surgery, or autopsy and dis- tinguished from helminths macroscopically or in tissue sections. In the liver (Fig. 8.13.7), intestinal wall, mesentery, mesenteric lymph nodes, peritoneum, or lung, viable encysted larvae, or granulomas containing necrotic pentastomes or their moulted cuticles may be found. Initially, encysted larvae excite little or no tissue reaction, but the granulomas are surrounded by hyalinized or calcified fibrous tissue. Serological tests are being developed, while a specific polymerase chain reaction that targets the 18S small subunit rRNA gene might be useful even in formalin-fixed paraffin-embedded tissue. Fig. 8.13.7 Encysted nymph/larva of Armillifer armillatus in human liver. The outer layer of the

parasite (arrowed) lines the cyst wall. Acidophilic glands (ag), intestine (in), ×21. Armed Forces Institute of Pathology photograph, negative number 75–2703.

section 8 Infectious diseases 1586 Treatment There is no specific treatment, although ivermectin, praziquantel, mebendazole, and albendazole have been used with apparent suc- cess. Obstruction and compression should be relieved surgically. Hypersensitivity phenomena should be treated with adrenaline (epi- nephrine), antihistamines, and corticosteroids. Prevention Pentostomiasis can be prevented by thoroughly cooking all meat of any origin and boiling or filtering drinking water. Eating sheep's lymph nodes is proscribed by the Shi'ite Muslims of Lebanon. Other zoonoses transmitted from reptiles to humans The most important of these is salmonellosis transmitted to humans by the faecal–oral route or by scratches and bites, from chelonians (tortoises, turtles, terrapins) and from snakes and lizards, espe- cially iguanas. In the United Kingdom, 38% of imported tortoises (Testudo spp.) contain salmonella. In the United States of America, where 8 million reptiles are kept as pets, contact with reptiles and amphibians accounts for an estimated 74 000 (6%) of the approxi- mately 1.2 million sporadic human salmonella infections that occur there annually. The banning by the United States Food and Drug Administration of commercial distribution of small turtles has prevented an estimated 100 000 cases of salmonellosis among chil- dren each year. Although salmonellosis usually causes self-limiting gastroenteritis, septicaemia or meningitis can occur especially in infants and immunocompromised people. Species associated with reptile salmonellosis include S. enterica serotype Typhimurium, S. enterica serotype Pomona, and S. enterica subspecies diarizonae. Other infections transmissible from reptiles to humans in- clude Arizona hinshawii (in snake powder, Pulvo de Vibora, made from rattlesnakes), Plesiomonas shigelloides, Edwardsiella tarda, leptospirosis, Q fever, sparganosis, capillariasis, strongyl- oidiasis, mesocestoidiasis, sarcocystiasis, and infestation with the mite Ophionyssus natricis. Potential zoonoses include mycobac- teria, pseudomonas, other aeromonas species, proteus, and some togaviruses (such as Western equine encephalitis in garter snakes in western North America) and herpesviruses. FURTHER READING Chen SH, et al. (2009). Multi-host model-based identification of Armillifer agkistrodontis (Pentastomida), a new zoonotic parasite from China. PLoS Negl Trop Dis, 4, e647. Lai C, et al. (2010). Imaging features of pediatric pentastomiasis infec- tion: a case report. Korean J Radiol, 11, 480–4. Lavrov DV, et al. (2004). Phylogenetic position of the pentastomida and (pan)crustacean relationships. Proc Biol Sci, 271, 537–44. Magnino S, et al. (2009). Biological risks associated with consumption of reptile products. Int J Food Microbiol, 134, 163–75. Palmer PES, Reeder MM (eds) (2001). Pentastomida. In: The imaging of tropical diseases with epidemiological, pathological and clinical cor- relation, Vol. 2, pp. 389–95. Springer, Berlin. Pantchev N, Tappe D (2011). Pentastomiasis and other parasitic zoonoses from reptiles and amphibians. Berl Munch Tierarztl Wochenschr, 124, 528–35. Riley J (1986). The biology of pentastomids. Adv Parasitol, 25, 45–128. Schacher JF, Khalil GM, Salman S (1965). A field study of Halzoun (parasitic pharyngitis) in Lebanon. J Trop Med Hyg, 68, 226–30. Sulyok M et al. (2014). Ocular pentastomiasis in the Democratic Republic of the Congo. PLoS Negl Trop Dis, 8, e3041. Tappe D, Büttner DW (2009). Diagnosis of human visceral pentastomiasis. PLoS Negl Trop Dis, 5, e320. Tappe D, et al. (2011). Diagnosis of human visceral pentastomiasis. Emerg Infect Dis, 17, 251–4. Tappe D, et al. (2011). Transmission of Armillifer armillatus ova at snake farm, The Gambia, West Africa. Emerg Infect Dis, 17, 251–4. Tappe D, et al. (2014). Imported Armillifer pentastomiasis: report of a symptomatic infection in the Netherlands and mini-review. Travel Med Infect Dis, 12, 129–33. Tappe D, et al. (2015). Molecular diagnosis of abdominal Armillifer grandis Pentastomiasis in the Democratic Republic of Congo. J Clin Microbiol, 53, 2362–4. Vanhecke C, Le-Gall P, Le Breton M, Malvy D (2016).

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