# Sudden death due to disseminated porocephalosis – a case history

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Received September 9, 1991 / Received in revised form November 27, 1991

**Summary.** An 18-year-old Nigerian girl died suddenly and unexpectedly from disseminated porocephalosis. The patient was hospitalised complaining of fever, dizziness, weakness and jaundice. Clinical examination revealed a restless, confused and hypotensive patient. She died within two hours of admission. Autopsy revealed disseminated Porocephalus armilatus infestation involving the thoracic and abdominal linings and their internal organs. The relationship between the disseminated parasitic involvement and the death of this girl is discussed.

**Key words:** Sudden death – Disseminated porocephalosis

**Zusammenfassung.** Ein 18 Jahre altes nigerianisches Mädchen starb plötzlich und unerwartet an einer disseminierten Porocephalose. Die Patientin war in das Krankenhaus eingeliefert worden und klagte über Fieber, Schwindel, Schwäche und Gelbsucht. Bei der klinischen Untersuchung fand sich eine unruhige, verwirrte Patientin mit niedrigem Blutdruck. Sie verstarb 2 Stunden nach der Einlieferung. Bei der Obduktion wurde ein disseminierter Befall mit Porocephalus armilatus festgestellt. Dieser betraf die Wände der Brust- und Bauchhöhle und deren innere Organe. Der Zusammenhang zwischen dem disseminierten parasitären Befall und dem Tod des Mädchens wird diskutiert.

Schlüsselwörter: Plötzlicher Tod – Disseminierte Porocephalose

## Introduction

Death from infection is commoner in developing countries than in industrial societies with better health care facilities. Such disease processes often manifest very characteristic and specific clinical symptoms and although death may result, it rarely occurs suddenly and unexpectedly. The forensic pathologist is therefore rarely concerned with such deaths.

revealed disseminated porocephalosis. Porocephalosis is a parasitic infestation caused by a

The case presented is of an 18-year-old girl who died

within two hours of hospital admission and the autopsy

worm-like arthropod of the genus Porocephalus and order Pentastomida and is closely related to another parasite of the genus Linguatula. Both are blood-sucking endoparasites of mammals, birds and reptiles [1, 2]. The adult form of *Porocephalus* inhabits the lungs of reptiles while the immature forms are found in intermediate hosts represented by many vertebrates including man [1, 2]. Infestation in man often leads to a "blind" end in the life cycle of the parasite and no inter-human transmission occurs. Porocephalus species are found in West, Central and East Africa, South-east Asia, China and Japan [3-7] and of approximately twenty species, P. armilatus and P. moniloformis are well known to cause infection in man in Africa and Asia respectively.

## Case history and autopsy findings

An 18-year-old Nigerian secondary school girl was admitted on 23 February 1989 into the Casualty Department of the Jos University Teaching Hospital in Nigeria complaining of fever, dizziness, jaundice and weakness. She had earlier taken a natural herbal concoction, routinely used in these areas to treat malaria which brought no relief. Examination revealed a restless and confused girl with a pulse rate of 136 per minute, blood pressure of 100/40 mmHg and a tender hepatomegaly. No definite diagnosis was made and before any further investigation could be carried out, she died about the hours following admission.

Autopsy revealed marked icteric discolouration of the sclerae. Widespread distribution of encysted P. armilatus nymphs were found on the entire peritoneal, pleural and pericardial surfaces, diaphragm, omentum, lungs, liver, serosal aspects of the uterus and pouch of Douglas and the para-aortic region (Figs. 1–3). The mucosal and serosal surfaces of the stomach and all the loops of the small and large intestines showed the presence of these parasites.

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Fig. 1. Liver, stomach and diaphragm (upper left hand corner) showing numerous encysted larvae

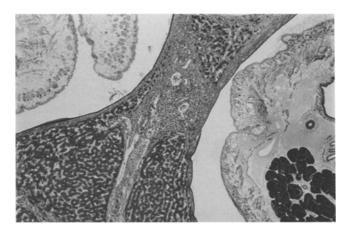


Fig. 2. Lungs and pericardial sac showing encysted larvae

Histology of the bowel wall revealed hyperplasia of the lymphoid follicles; encysted nymphs were identifiable within all the layers of the bowel wall and were surrounded by fibrous tissue. The omentum and mesentery showed a similar picture with moderate local eosinophil



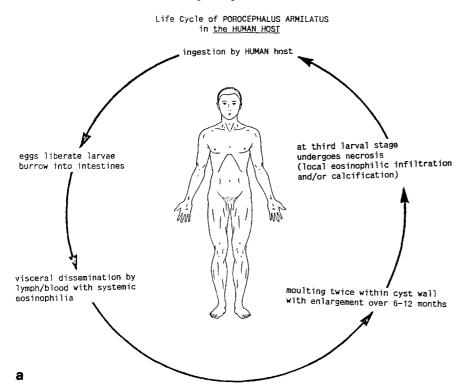
Fig. 3. Para-aortic tissue, serosal aspects of urinary bladder and uterus showing numerous encysted larvae



**Fig. 4.** Encysted *P. armilatus* larva within the liver parenchyma (H & E  $\times$  60)

infiltration. The hyperplastic mesenteric nodes exhibited sinus histocytosis.

Macroscopical sectioning of the liver, which weighed 2500 g, showed that over 80% of its entire parenchyma was infected by the nymphs. Histology showed minimal



Life Cycle of POROCEPHALUS ARMILATUS in the <u>animal reservoir</u>

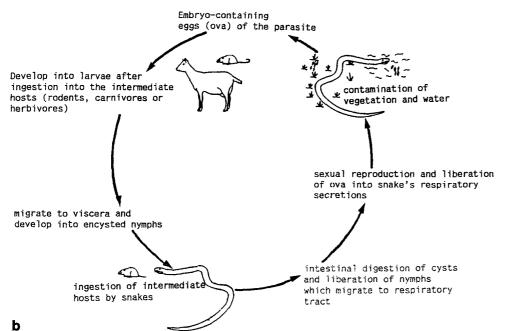


Fig. 5a, b. Life cycle of *P. ar-milatus* in animal reservoir and man

cellular inflammatory response around the cyst walls (Fig. 4); the inflammatory cells included lymphocytes, plasma cells, eosinophils and some histiocytes. The hyperplastic Kupffer cells contained malarial pigments.

The spleen, which weighed 250 g, had a purplish colour and a slightly firm consistency. The cut section showed a dark red surface, well defined margins and mild to moderate softening of the pulp. Histology revealed marked

congestion of the red pulp with relative 'atrophy' of the white pulp.

On sectioning the lungs showed numerous encysted nymphs with a paraseptal emphasis and mild pulmonary oedema.

Many encysted nymphs were also identified within the rectus abdominis muscle, diaphragm and the intercostal muscles. The intravascular compartments of most organs including the brain contained numerous eosinophils.

Death was attributed to disseminated porocephalosis. No facilities for toxicological studies were available but extensive inquiries among the next of kin and the school friends of the deceased failed to reveal any indications of poisoning.

### Discussion

The adult forms of *P. armilatus* are found in the lungs, trachea and nares of pythons, giant vipers, cobras, mambas and puff adders [1-4]. The ova of the parasite are shed with the bronchial secretions of these animals and remain viable in water or soil for up to six months. Man acquires the disease after ingesting contaminated water, and uncooked or improperly cooked vegetables contaminated by snake secretions. Infection also results from eating improperly cooked snake meat or flesh of other infected intermediate hosts such as rodents, wild and domesticated animals (Fig. 5a and b). The larvae (measuring  $92.0 \times 72.0$  microns) are released into the human small intestine. Each has 4 legs terminating in a pair of claws and an anterior penetrating apparatus with which it pierces the intestinal wall. The larvae are carried by blood or lymph to the liver and other organs where they encyst and lose the penetrating apparatus and claws. Moulting occurs twice and the third (much larger) larval stage which is usually reached in one year, consists of a full grown, closely coiled, encysted nymph that resembles the adult. No further development occurs in man and the disease generally remains symptomless.

There is no information as to the pathology of the 'migratory phase' in man and no toxins derived from the parasite have been isolated [4]. Evidence of infestation in man is mostly detected by radiographs showing calcified nymphs within the chest and abdomen. Linder (1965) reported on 1000 randomly selected abdominal X-ray films in Ibadan, (South West Nigeria) and found that 1.4% of the films showed calcified nymphs of *Porocephalus*, with the right upper quadrant being the commonest site [5]. An autopsy series in Ibadan showed that 2% and 4% of male and females respectively had encysted nymphs [6] and reports from Congo, Cameroun and Malaysia revealed infection rates of 27%, 6–13% and 45% respectively [3].

It appears that porocephalosis is mainly seen among the Yorubas of South West Nigeria [4–9]. Although the deceased was resident in Central Nigeria she was of the Yoruba tribe. Her parents denied any family tradition of eating snakes, but her school colleagues confirmed that they frequented possibly contaminated streams to fetch water for daily use.

There have been no previous reports of sudden death in association with simple porocephalosis. The disease is known to follow a chronic course and the severity of clinical manifestations depends on the intensity of infection. There is no known treatment and prognosis is believed to be poor if the nymphs escape from their cysts and migrate into the tissues [1]. Multiple infections have resulted in severe abdominal pain and icterus, as was observed in the deceased, when the larvae migrate from the intestine to other organs. Pneumonitis, bronchitis, hepatitis and menigitis have been reported [1, 4]. Obstructive jaundice [2], hepatic venous occlusion [8] and intestinal obstruction [10, 11] have also been documented.

The present case in unique in that the only condition found at autopsy was the overwhelming parasitic burden which appears to have produced a state of shock. Although facilities for toxicological analysis of the native concoction were unavailabe, poisoning was not considered a possible cause of death. This is because the concoction, a common traditional medication, had been used on earlier occasions by the deceased and is very commonly used elsewhere in the country with no known ill-effects.

Acknowledgements. The staff of the Medical Illustration Department, University of Jos Medical School produced the gross photographs; the staff of the Histopathology Laboratory, Jos University Teaching Hospital prepared the slides and the Photography Sections, Forensic Medicine Unit, University of Edinburgh Medical School produced the microphotographs and drawings. Mrs. Joan Bell deligently typed the manuscripts. The authors are grateful to them all.

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