Cranial and Intracranial Hydatidosis

With Special Reference to Roentgen-Ray Diagnosis

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For many years emphasis has been laid upon the fact that tapping an intracerebral hydatid cyst or tearing the membrane of a cyst during an operation is extremely detrimental.\textsuperscript{1,2,13,17,18} The spilling of fertile cysts into the surrounding brain substance and into the subarachnoid space enhances the danger of recurrence and meningeal symptoms both of which are serious complications. An exact diagnosis should be aimed at not only in the localization of the lesion but also in the determination of its nature. In this report some of the neuroradiological peculiarities in 28 consecutive cases of cranial and intracranial hydatidosis are discussed and the corresponding literature is reviewed.

Extracerebral Hydatidosis

1. Cranial Form. Forty years ago Dévé\textsuperscript{6} injected scolices into the carotid artery in rabbits and succeeded in producing hydatid cysts in several bones. The scapula, the bony structure of which resembles the skull in many respects, was involved in one of these animals. Study of the specimen of bone revealed that the spongiosa was the first part to be involved. Furthermore it was noticed that the pattern of multiplication of cysts within the bony substance was determined mainly by the pressure exerted by the tissue of the host which resists any further extension of the lesion. This kind of development was termed by Dew\textsuperscript{8} as “exogenous vesiculation.” The resulting cystic material is a pulpy mass consisting of numerous microcysts and their necrotic remnants. With further progression of the lesion the compact tissue is bulged and perforated at many spots, allowing the cysts to invade the soft tissue, where they may grow to the same size and shape as encountered in parenchymatous organs. The pathology of echinococcosis involving the calvarium corresponds to a high degree to that in animals.

Case 1. Five years prior to admission the relatives of a 10-year-old girl noticed a bulging of the right side of her head. This bulging slowly increased in size and induced the parents to consult our clinic. Examination revealed a very big prominence of the right side of the head (Fig. 1). It felt as hard as normal bone, and pressure exerted upon it caused no pain to the patient. No signs indicating intracranial hypertension and no neurological abnormalities could be detected. Roentgenograms of the chest were normal, Casoni’s and Weinberg’s reactions were negative, and no eosinophilia was found in the blood. Plain films of the skull revealed an enormous bulging of the external table, though the internal table appeared to be thin and slightly impressed. The space between the two tables included a great number of thin bony laminae, leaving between them numerous cavities (Fig. 2). A diagnosis of bone tumour of unknown nature was made.

Operation. In moderate hypothermia a skin flap was turned. Burr holes were made 1 cm.

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away from the border of the tumour and connected with Gigli saws. The dura mater was separated carefully from the internal table and the growth was removed in one piece. The patient tolerated the procedure without any complication. Plastic repair of the bony defect has not yet been performed.

The specimen weighed 430 gm. It had almost the shape of a hemisphere in which a paper-thin internal table formed the basis. Upon its removal a great number of cavities became visible which were separated by thin laminae of bone. Most of these cavities were filled with necrotic cysts; others were empty. The external table, which formed the dome of the hemisphere, presented a normal configuration.

Comment. Exclusive localization of hydatid cysts within the skull seems to be a very rare occurrence, if one realizes that in man only 2 per cent of hydatid cysts are localized in the skeleton, and of these only 3.4 per cent are in the skull.6 In his interesting book, Dévé could collect from the literature until 1948 not more than 23 instances of cranial hydatidosis. Unfortunately he did not mention in how many of these patients the skull was involved solely. Larger series of cerebral hydatidosis, published in recent years, include only exceptional cases with cranial involvement.4,19,20 Goinard et al.10,11 reported on a 21-year-old patient in whom plain films of the skull demonstrated, in addition to numerous bony defects of the vault, two linear calcifications within the cranium, which ran almost parallel to each other. The nature of these lines was identified during the operation; one was the internal table pushed into the cranial cavity by numerous cysts and the other was the underlying calcified dura mater.

2. Cranial and Extracranial Form.

Case 2.* A 23-year-old officer was admitted with the chief complaint of headaches and dizziness. In the last 12 months he noticed numerous swellings in both occipital regions of his head. Two months before admission the patient started to have headaches and vomited on several occasions. He felt somewhat dizzy and tired, could not concentrate and was unable to attend his classes. Clinically we detected in both parietal and occipital regions at least five fluctuating swellings, the biggest one being 1.5 cm. prominent with a diameter of 4 cm. Bilateral papilledema was noted. The gait was unsteady but not truly ataxic. He was slow in thinking and inattentive in conversations. Plain films of the skull (Fig. 3) revealed in the parietal and occipital regions large bony defects separated from each other by thin laminae of bone. Both internal and external tables were irregularly destroyed and showed at several points the circular outlines of small hydatid cysts. The probable diagnosis of cranial echinococcosis was made.

Operation. Following the skin incision, cysts became visible in the subgaleal space. Through the largest bony defect the intracranial space was explored. It was filled with hundreds of cysts which were removed. The thinned dura mater was seen to be pushed to the bottom of the cavity, which occupied approximately one-third of the whole intracranial space.

Course. Complete recovery followed the operation. One year later, however, the patient consulted us for a fistula from which infected cystic material was still escaping.

Case 3. A 32-year-old farmer consulted us for a swelling on his right forehead.

Examination. There was a soft and moderately painful swelling with a prominence of 2-3 cm. and a diameter of 6 cm. No neurological deficits were found. On plain roentgenograms (Fig. 4) a large and well-limited bony defect in the right frontal area became visible. The edges were sharply defined and consisted of a rather con-

* This case has been published previously by one of the authors.20 We are grateful to G. Thieme Verlag for permitting us to print this translation.
3. Combined Form. By this term we mean those patients in whom simultaneous intracerebral, extradural and bony lesions are found.

Case 4. A 17-year-old boy was admitted with a history of 5 months' duration. He complained of headaches, vomiting, diplopia, and disturbance of speech.

Examination. He presented bilateral papilledema, right facial weakness, some mental deterioration and motor dysphasia. Left carotid arteriography (Fig. 5) revealed a huge space-occupying mass within the frontal lobe. The overlying bone showed a localized area of destruction. Operation. During the craniotomy the bone flap showed a limited zone of necrosis of bone. Extradurally a number of cysts were found and removed. Upon opening the dura mater the bluish membrane of an intracerebral cyst became visible. It was carefully dissected from the surrounding brain and removed without being ruptured.

Postoperative course was uneventful.

Comment. Even in such a case with an extensive intracerebral lesion, the origin of the disease remains obscure. Gripponissiotis attributed this combination to injury which may cause a rupture of the primary intracerebral cyst attached to the dura mater. Goinard et al. reported a case with erosion of the temporal bone by underlying extradural cysts. The intracerebral lesion was so extensive that it even reached the wall of the lateral ventricle. A remarkable case which belongs to this category of pathology was reported by Krebs et al. Their 28-year-

Fig. 3. Case 2. Roentgenogram showing extensive involvement of the extradural space in cranial hydatidosis.

densed bony material. These findings were compatible with an epidermoid of the skull.

Operation. The skin was incised around the defect. During its dissection the thin membrane of a solitary hydatid cyst ruptured and colourless fluid escaped. The inner membrane of the cyst was then removed. The dura mater was not involved and we had no reason to explore the intradural structures.

Course. The postoperative period was uneventful.

Comment. The existence of primary extradural echinococcosis is still a matter of question. Extradural vessels which may carry hexacanth embryos to this space are few or nonexistent. In one of our cases with normal roentgenograms of the skull circumscribed erosions of the internal table were found during the craniotomy. We interpreted them as primary lesions leading to the development of numerous extradural cysts. On the other hand the extradural space may be infested by intracerebral cysts through an apparently healthy dura mater. In our two cases the bony lesion was so widespread that we had no doubt of its being the primary lesion. The cavity filled with cysts may assume huge dimensions as in our Case 2. Surprisingly, the physical disability produced is not in keeping with the extent of the lesion. One should realize however that a great deal of the pressure is transmitted extracranially through numerous defects in the bone.

Fig. 4. Case 3. Roentgenogram showing defect of bone caused by a solitary hydatid cyst.
old patient presented an involvement of cranial nerves IX, X, XI, and XII and a soft swelling behind the ear. He complained of headaches, vomiting and vertigo. Following his sudden death, the postmortem examination revealed 10 extradural and 10 intradural cysts. The bone covering the ipsilateral posterior fossa was found to be necrotic.

Intracerebral Hydatidosis

1. Effects of General Pressure. Effects of pressure on the brain manifest themselves within the first or second decade of life with a predominance between 6 and 18 years. Though Dew’s statement that “a hydatid cyst is just about as old as the patient” is somewhat exaggerated, the length of history does not represent the length of time the cyst has been present. Some very big cysts have been recorded and it is a matter of surprise how the intracranial contents adapt themselves with so few pathological changes and so few clinical effects. With such a chronicity in mind, it is evident that straight roentgenograms of the skull show the effect of a long-standing intracranial hypertension. Increase in convolutional markings, opened sutures, enlargement of the sella turcica with erosion of its apophysis are seen as a rule. They have no relevance to the specific diagnosis of cerebral hydatid cysts.

2. Effects of Local Pressure. Localized bulging of the skull is an indication of the length of time the lesion may have been present.

Case 5. A 9-year-old girl was admitted complaining of headaches, intermittent vomiting and impairment of vision. The history of the illness could be followed up to 2 years before admission when she started to have headaches. A decrease in visual acuity was noticed 3 months prior to admission and its total loss 2 months later.

Examination detected a bilateral papilledema with consecutive atrophy of the optic nerve. Ventriculography, during which a cyst was inadvertently tapped, revealed a displacement of the ventricular system. The bone overlying the cyst showed a local bulging (Fig. 6).

Operation. A left frontal craniotomy was performed. Three multiloculated cysts were removed.

Course. A cerebrospinal-fluid fistula with subsequent purulent meningitis developed. The child
died 3 months later in a state of decerebrate rigidity.

Comment. Local bulging of the skull is met with in a number of superficially situated tumours or collections of fluid. In cases of hydatid cysts the bone may become very thin so that crepitation may be felt with the finger.\textsuperscript{11,21,23} Tangential projections are of great value in demonstrating these changes, which should not be confused with bony defects caused by direct involvement of bone.

3. Linear Calcification. As a rule there is very little reaction in the glial tissue surrounding a cyst, so that typically the adventitious capsule is ill-defined or practically absent. A linear calcification of the adventitious capsule was unknown until 1944, when the first instance was recorded by Kooy.\textsuperscript{15} Its frequency probably does not exceed 1 per cent of all cases.\textsuperscript{21}

Case 6. An 18-year-old boy was admitted for headaches, intermittent vomiting, and general epileptic attacks which started 6 months previously.

Examination revealed bilateral papilledema and discrete motor weakness on the left side. Straight roentgenograms of the skull (Fig. 7) revealed a tiny linear calcification with anterior concavity, beginning in the frontal vault and ending in the base near the sella. In our opinion this finding was compatible with the diagnosis of a cerebral hydatid cyst. Subsequent carotid arteriography showed a huge avascular mass corresponding to the aforementioned circular line.

Operation. The cyst was approached through a frontal craniotomy. It was covered by a thin layer of cortex which was incised and the cyst was removed unruptured. The adventitious tissue was surprisingly tough and contained islands of calcareous deposits. It was left untouched.

Postoperative course was uneventful.

Comment. In Dew's\textsuperscript{8} case of an 8-year-old female, in whom a linear calcification outlined the cyst, the adventitia was found to be very thick and irregularly calcified. This "rigid, not collapsible calcareous wall, not unlike a thick eggshell" was not removed. The patient described by Kooy\textsuperscript{16} was a 27-year-old man suffering from headaches since the age of 8 years. One was struck by an asymmetry of the face and the skull, of which the right half showed a considerable bulging. The left side of the body was less developed. Plain roentgenograms of the skull showed marked asymmetry, no thickening or thinning, but a large linear calcification through the hemisphere. During the operation one found a colossal cavity filled with a conglomeration of cysts and caseous matter, the cavity occupying the complete right half of the skull.

4. Circumscribed Calcification. The calcification within a cerebral hydatid mass does not necessarily represent the outlines and the nature of the lesion. It is irregular, wedge-shaped and nonhomogeneous in intensity. Even during the operation a tuberculoma can not be discarded with certainty unless necrotic membranes are identified. In some instances, however, the calcification is so well outlined that a hydatid lesion may be determined with certainty.\textsuperscript{18}

Case 7. A 27-year-old female consulted us mainly because of her ataxic gait. At the age of 14 she had an episode of headaches, vomiting, some impairment of vision and severe ataxia. This condition was then diagnosed as being an encephalitis and treated as such. She recovered slowly and incompletely.

Examination. When admitted, she displayed a

![Fig. 7. Case 6. Roentgenogram revealing linear calcification (arrows) representing the calcified adventitious membrane of a solitary intracerebral cyst.](image-url)
5. Cystography. The tapping of a cyst and replacing its contents with air was the method of diagnosis in the area when cerebral angiography was not yet popular. In patients in whom ventriculography is performed, the cyst may be tapped in 40 per cent of the cases according to Obrador.\textsuperscript{19} Subsequent contamination of cerebral tissue with fertile scolices or invasion of cerebrospinal-fluid pathways with parasites may become fatal.\textsuperscript{13,14,19} Nowadays cystography should never be performed intentionally.\textsuperscript{1,2,8,17}

**Case 8.** A 7-year-old boy was admitted because of chronic headaches and occasional vomiting.

*Examination* revealed a head larger than normal, bilateral papilledema and moderate weakness of the right extremities. During ventriculography a solitary cyst was tapped. Subsequent radiography (Fig. 9) revealed a big hydatid cyst of the parietal region.

*Operation.* Attempt was made not to rupture the membrane of the cyst. Unfortunately an enlargement of the hole made by the brain cannula could not be avoided, so that the whole field became probably contaminated with fertile scolices.

*Course.* From the 3rd postoperative day on, a pyrexic condition with severe meningeal symptoms developed. Analysis of the cerebrospinal fluid, obtained by lumbar tapping, revealed 63 mg per cent protein, 180 lymphocytes and 9 leucocytes in mm.\textsuperscript{3} His condition deteriorated rapidly and he expired 7 days after operation.

*Comment.* The appearance in cystograms of a fold of membrane across the cyst gives
an appearance very suggestive of what in pulmonary cysts has been called the sign of Calotte. Sometimes an irregularity in the outline of the cyst may simulate the mural nodule of an astrocytoma. If the air escaping from puncture of the cyst passes into the space between it and the brain, the membrane of the cyst becomes visible and the diagnosis can be made with certainty. Should the cyst contain daughter cysts, they are seen as small spherules on the inner side of the mother cyst, as Arseni and Samitca observed in 10 per cent of their cystograms. These authors stated also that sometimes the daughter cysts float freely within the mother cyst. The simultaneous filling of multiple exogenous daughter cysts of fair size may develop from sclices shed through a small hole in the laminated membrane of the original mother cyst. We saw also 2 instances in which a number of cysts could be identified on the cystogram.

6. Ventriculography. Ventriculograms in cases of hemispheric cysts offer no peculiar pattern that would allow a specific diagnosis. The ventricular system opposite the lesion is usually enlarged, the homolateral ventricle being thinned to a line or even being absent. Because of its inherent hazards, ventriculography is being more and more replaced by angiography. Nevertheless, there are cases of cerebral hydatidosis in which ventriculography seems to be the ideal method of diagnosis. Symptoms such as spastic tetraparesis or decerebrate rigidity, simulating a midline lesion or a tumour somewhere in the posterior fossa, still require ventriculography. In cases of para-ventricular cystic formation angiography is of little value. The same is true when the cyst is lying completely within the 3rd ventricle or in the posterior fossa. If a hydatid cyst is suspected and ventriculography seems to be unavoidable, it is then advisable to puncture the ventricle opposite to the suspected cyst or discontinue the procedure as soon as the top of the cannula meets the resistance of the membrane of a cyst.

7. Cerebral Angiography. We can only reaffirm the statement of previous observers that this technique is of relevant value in the determination of cerebral hydatidosis. A number of angiographic peculiarities are outlined sketchily by Arana Iniguez et al. They are: (a) Paucity or even absence of vessels within a surprisingly large area not in keeping with disabilities produced. (b) Circumferential straightening of the vessels which outline a spherical mass. These vessels are arranged parallel along the surface of the cyst. (c) The arteries have an even diameter throughout their course. They are not tortuous, dilated or narrowed.

Case 9. A 12-year-old boy was admitted mainly because of impairment of vision in both eyes. During the last 4 months prior to admission he had periods of severe cephalaea. Examination revealed advanced papillledema with beginning consecutive atrophy of the optic nerve. The only detectable focal sign was a facial weakness on the left side. Left carotid angiography was done (Fig. 10). The anteroposterior view showed an extensive displacement of the an-
terior cerebral artery, which was circularly arranged and outlined an avascular area within the frontal region.

Operation. A large cyst situated subcortically was identified and removed without being ruptured.

Course. Examination 4 months after operation revealed great improvement of sight in both eyes.

Comment. We subjected 20 patients with mainly supratentorial hydatid cysts to carotid angiography. In 7 cases a tumour was suspected but cysts were found and removed surgically. In 13 patients the angiographic diagnosis of hydatid cysts was confirmed during the subsequent operation. In 9 patients in whom clinical data including cerebral angiography were in favour of hydatid disease the exploration disclosed the true nature of the lesion, such as intraventricular meningiomas, epidermoids, tuberculomas, cystic gliomas and abscesses. In adults the discrepancies seem to be still more pronounced. Ten adult patients, harboring cerebral hydatid cysts, were subjected to angiography by Arana Tiniguez and Maslenikov. In only 3 of them did the angiographic pattern fulfill the conditions required by these authors; in the rest the diagnosis was that of a tumour.

Discussion

The incidence of pure cranial hydatidosis is so rare that its diagnosis still remains a matter of speculation (Case 1). The pathogenesis of this condition is one of ischemic necrosis within the spongiosa with subsequent bulging of the internal or external table. A “fenestration” of these structures allows the cystic material to escape into the extradural space entailing a displacement of the brain, whereas an outward extension manifests itself in numerous subgaleal swellings (Cases 2 and 3). The origin of the pure extradural collection of cysts without involvement of the bone or underlying structures still has remained unknown. In cases of intracerebral cysts with subsequent contamination of the extradural space and erosion of the neighbouring bone, the diagnosis is feasible by straight roentgenograms and cerebral angiography (Case 4). In most cases, however, the hydatid cysts are located within one cerebral hemisphere.

Their occurrence in children and the long-standing compression of the brain entail secondary effects of pressure such as open sutures, changes in the sella and increase in convolutional markings. Localized effects of pressure, such as bulging or thinning of the skull, brought about by an underlying cyst (Case 5) have been produced experimentally by Dévé. Roentgen-ray signs indicating general or local effects of pressure are certainly not specific for hydatid disease. Linear calcification of the adventitious tissue surrounding a cyst is a very exceptional finding. Its presence is of great diagnostic value, as it outlines the limits of a cyst (Case 6). Less characteristic is a calcification within a chronic hydatid conglomerate (Case 7). Spherical calcifications however, such as published by Legré and Massad, leave no doubt about the nature of the lesion in question. Ventriculography, so strongly advocated by former authors, is being abandoned more and more because of its hazards. It is still the method of choice in patients presenting the symptoms of a midline lesion. The same can be said of cystography which was at times a popular method of diagnosis. Spilling of fertile scolices with subsequent contamination of the neighbouring brain tissue and the cerebrospinal-fluid pathways are reasons why this procedure has lost its former value.

At present cerebral angiography seems to be the most reliable method in localization of hydatid cysts and in determination of their nature. Avascularity of the lesion and circular arrangement of vessels surrounding a cyst are almost specific (Case 9). This seems not to be true in adult patients in whom the nature of the lesion can not be identified with certainty.

Summary

Twenty-eight consecutive cases of cranial and intracerebral hydatidosis were analysed mainly from point of view of roentgen-ray diagnosis. Changes on straight roentgeno-
grams of the skull are discussed and evaluated. Ventriculography with inadvertent injection of the cyst with air (cystography) is being replaced more and more by cerebral angiography. Angiography has a far-reaching diagnostic value in localization of hydatid cysts and determination of their nature.

References