Neurenteric Cyst Diagnosed by Technetium-99m Pertechnetate Sequential Scintigraphy

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Neurenteric cysts are rare congenital anomalies which present as mediastinal tumors associated with vertebra anomalies. Two-thirds of them are lined with gastric mucosa and are potentially life threatening. An exact differential diagnosis is difficult preoperatively but is absolutely necessary because of the grave prognosis if left untreated. We present a case in which [^{99m}Tc]pertechnetate sequential scintigraphy demonstrated gastric mucosa in the cyst and helped to confirm the diagnosis. The scintigraphic findings are correlated with radiologic, sonographic, and pathologic features.

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Neurenteric cysts are rare tumors of the posterior mediastinum. They are partially or completely lined by gastric mucosa and are associated with congenital anomalies of the vertebrae. Fistulae or solid connections to the central nervous system (CNS) are possible, and therefore they are ascribed to the split notochord syndrome. An early preoperative diagnosis is absolutely necessary because of the complications of such cysts which lead to a grave prognosis. Such complications are first due to acid and pepsin-containing secretions of the gastric mucosa in the cyst which lead to ulceration and perforation, second, to CNS infections if there is a fistula between the cyst and the spinal canal. For the purpose of an early diagnosis, technetium-99m (^{99m}Tc)pertechnetate scintigraphy can be used. This is a simple, noninvasive technique and can be applied without hazard to the patient. It demonstrates the ectopic gastric mucosa of the cyst and is therefore helpful in differential diagnosis.

CASE REPORT

A 6-wk-old female infant developed meningitis and was admitted to our institution. A chest radiograph and spot films showed hemivertebrae of the second and third thoracic vertebra with partial fusion. No other anomalies were established. At 4½ mo of age the patient again developed meningitis and an ampicillin-sensitive enterococcus was detected. The chest roentgenograph showed in addition to the known vertebra anomalies a right-sided mediastinal mass from T2 to T8 (Fig. 1). Because of normal urine catecholamines the differential diagnosis of a neuroblastoma was ruled out.

The patient often passed black, tarry stools. Because of the combination of abnormalities of the vertebrae, mediastinal tumor, and passage of black tarry stools a neurenteric cyst with intermittent gastric mucosa bleeding was suspected.

Because of the meningitis a fistula between the cyst and the spinal canal was suspected, but it was not confirmed by myelography. The upper gastrointestinal series showed absence of rotation of the colon and a small tumor with a central recession on the back wall of the duodenal bulb which impressed gastroscopically as a papillary bulging of the mucosa. This finding suggested, that this was the point of connection of a second fistula to the GI tract. The computer tomogram showed the known myelodysplasia and the tumor of the posterior mediastinum with soft-tissue density (Fig. 2). A cyst could not be diagnosed with certainty. The cyst was not visualized by sonography.

To confirm the diagnosis of an neurenteric cyst sequential $^{99m}TcO_4$ scintigraphy was performed. Imaging was done with a large field-of-view (LFOV) gamma camera* using a sequential technique. Immediately after i.v. injection of 2 mCi (74 MBq) [^{99m}Tc]pertechnetate the first picture was obtained, followed by images every 5 min up to 40 min after injection. The infant was sedated and neither perchlorate nor stimulating agents such as pentagastrin were administered prior to the examination.

The scintigraphy revealed an increasing uptake in the suspected cyst 5 min to 40 min p.i. without change of the shape of the scintigraphic finding (Fig. 3).

The patient later developed hydrocephalus malresorptivus caused by the meningitis. The hydrocephalus was treated by a shunt. After recovery, the mediastinal tumor was operated. Intraoperatively a fistula to the spine was found and was

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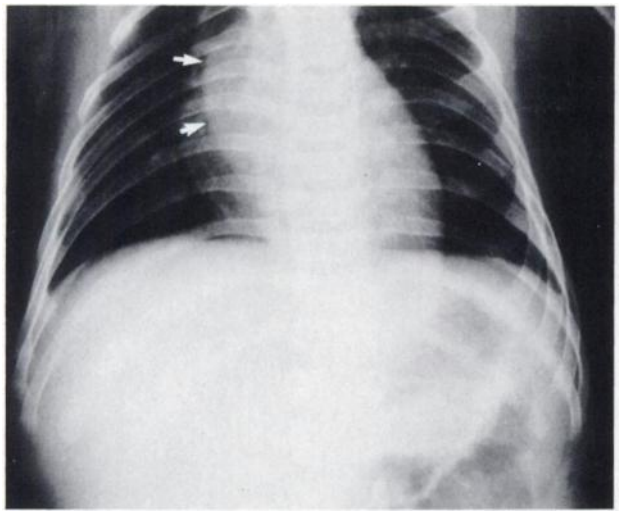

FIGURE 1
Chest radiograph at 4½ mo of age shows right sided thoracical mass (arrows).

disconnected from the vertebrae as close as possible. From the lower pole of the cyst a second fistula passed just beside the vena cava inferior through the diaphragm to the basis of the duodenum. After a short cut in the upper gastrointestinal region, the cyst and the fistulae were completely resected.

The histologic examination of the cyst demonstrated that the wall consisted of gastric and foregut mucosa tissue. A peptic ulcer was not established.

The postoperative course was free of complications and the infant is now 1 yr old and in good health.

DISCUSSION

Neurenteric cysts in the definition of Neuhauser et al. (*1*) are rare cases of intrathoracic masses. Only 73 cases are found sufficiently documented in the literature. The pathogenesis is extensively described by Koester (*2*) in his comprehensive review of the literature and the various connections of the cysts to the CNS and GI tract by Bentley (*3*). These cysts develop if the notochord and foregut failed to separate in the normal way during embryonic life.

An early diagnosis is absolutely necessary because of the invariably fatal outcome without adequate treatment—out of the 73 all 14 patients without an operation died—and there is an excellent prognosis after prompt surgery. There are, however, many diagnostic problems which may hamper the proper diagnosis, particularly when the cyst is ruptured.

The detection of ectopic gastric mucosa using [^{99m}Tc]pertechnetate scintigraphy is well established (*4–7*). The method has been used, for example, to detect gastric mucosa in Meckel's diverticulum (*8*) or in the Barrett's esophagus (*9*). There are, however, only few reports on the detection of neurenteric cysts (*10–12*). With regard to the [^{99m}Tc]pertechnetate scan it has to be considered that ^{99m}Tc uptake normally occurs in the salivary glands, thyroid (orthotopic or ectopic), and stomach. Pathologic uptake has been established in neurogenic tumors (*13*), vascular disorders (*14*), and in hiatus herniae associated with a short esophagus (*12*). On the other hand, one must keep in mind that only two-thirds of neurenteric cysts are lined with gastric mucosa.

Differential diagnosis can be facilitated by adequate scanning projections, sequential technique (*15*), and eventually by an additional use of ^{123}I-scintigraphy to exclude ectopic thyroid tissue.

In our case, only one projection was feasible because of clinical condition of the patient and the confirmed radiological localization of the mediastinal tumor. We

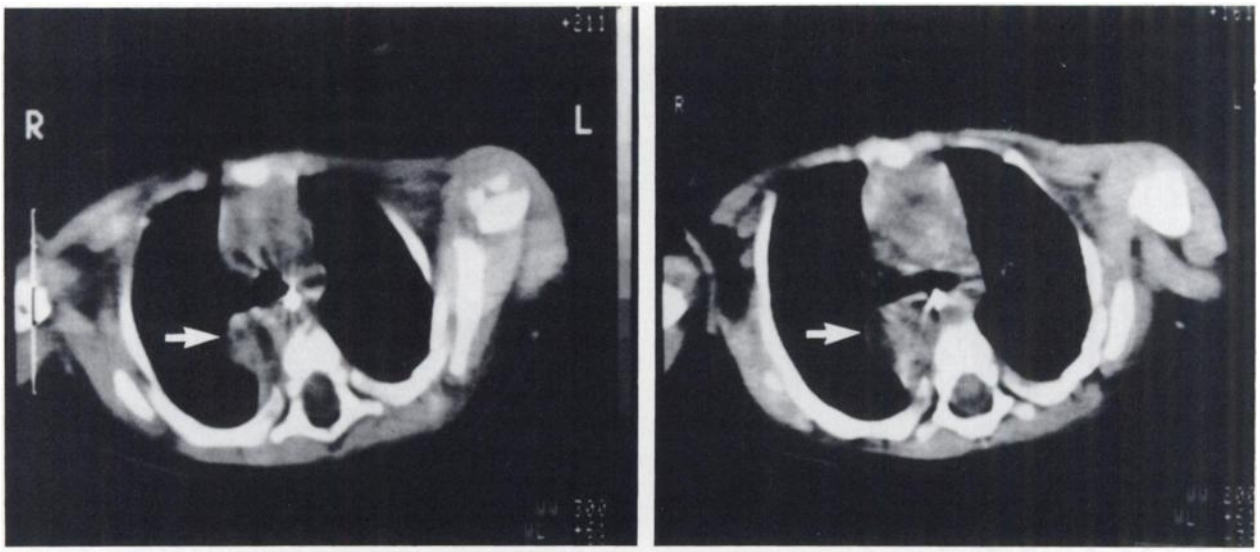

FIGURE 2
Two slides of the CT scan show the right sided tumor in the posterior mediastinum (arrows).

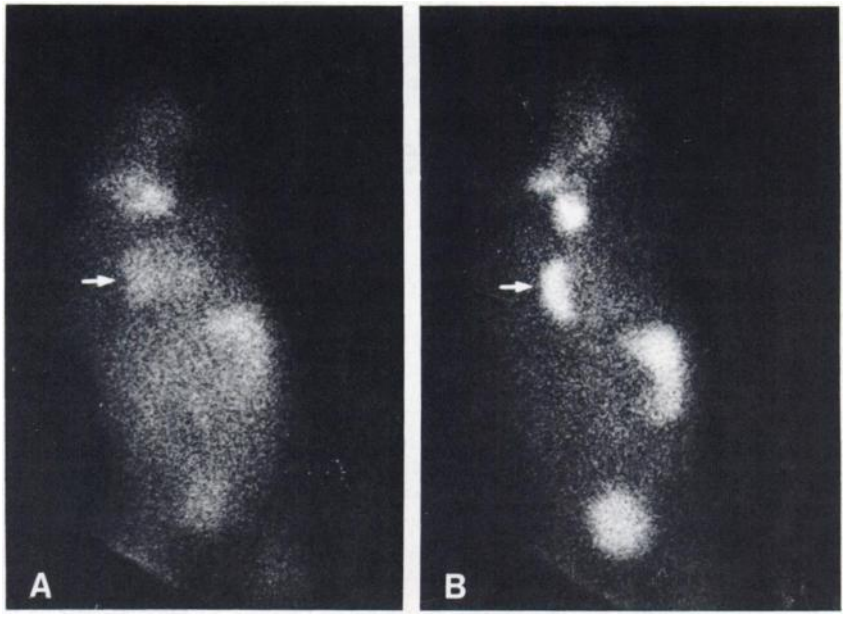

FIGURE 3
[^{99m}Tc]pertechnetate scintigraphy in anterior view shows pathologic accumulation of the radioactive tracer in projection to the tumor (arrows). A:5 min after injection. B:40 min after injection.

also did not perform ^{123}I-scintigraphy, as ectopic thyroid tissue was very unlikely because the tumor laid in the posterior mediastinum.

Barrett's epithelium which normally occurs in the lower part of the esophagus could be excluded with a high probability by reason of the shape and localization of the scintigraphic finding.

In conclusion, as tumors of the posterior mediastinum associated with vertebral anomalies are suggestive of neurenteric cyst it is advisable to perform sequential [^{99m}Tc]pertechnetate scintigraphy which can lead to the correct diagnosis in connection with the clinical history and radiologic findings.

NOTE

* (Sigma 410) Technicare, Solon, OH.

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