Desmoid Tumor Arising around the Distal Tubing of a Cerebrospinal Fluid Shunt

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A case of desmoid tumor in the abdominal wall as a cause of malfunction of a cerebrospinal fluid shunt is presented. The desmoid tumor arose from the reactive fibrose tissue formed around the silastic distal tubing and caused the catheter to become disconnected from the reservoir.

KEY WORDS: Hydrocephalus; Cerebrospinal fluid shunt; Desmoid tumor; Silicone implants

Although the shunting of ventricular fluid is the preferred treatment for hydrocephalus in children, there is no doubt that the surgical procedure itself and the shunt mechanism cause numerous complications. This clinical report presents the exceptional case of a child with a cerebrospinal fluid (CSF) ventriculoperitoneal shunt in whom the formation of a desmoid tumor within the abdominal wall around the peritoneal tubing caused malfunction of the system.

Case Report

This 19-month-old child, suffering from congenital hydrocephalus due to a Dandy–Walker malformation, had undergone a ventriculoperitoneal shunting of CSF performed at birth. The parents sought medical attention on October 19, 1982, because the child presented with two tumors. The first one was subjacent to the operative scar in the hypochondrium and had increased in size in the preceding 6 months. When the tumor was first detected, plain anteroposterior and lateral roentgenograms of the skull, chest, and abdomen showed normal results. Further examination revealed a hard, painless mass 6 × 8 cm in size that was adherent to the abdominal muscles. The second mass, located in the retroauricular area, suggested a fluid content. General examination of the child proved normal. X-ray study of the chest and abdomen showed the distal tubing of the shunt unattached and free in the peritoneal cavity, and radiologic signs of a mass in the right hypochondrium wall. There were no calcifications or lesions of the rib cage.

On October 21, 1982, the patient was operated upon with total removal of a tumor 6 × 4 × 2 cm in size, encapsulated and clearly delineated within the muscle of the abdominal wall. The tumor extended cranially along the costal wall with a subcutaneous fibrous tract. A further prolongation extended to the peritoneum and left a large defect in it when the mass was removed (Figures 1 and 2). The retroauricular mass was a subgaleal accumulation of cerebrospinal fluid. The wounds were closed in layers after the placement of a new peritoneal catheter.

The tumor mass when removed was firm and when cut was pearly white in color. Histologic examination showed it to be a neoformation of marked desmoplasmic characteristics, with fascicular formations of mature and atypical fibroblasts that infiltrated the muscle. It contained frequent mitotic figures (Figure 3).

The postoperative course was normal. Clinical examinations, abdominal and cranial computed tomography, and echography of the abdomen performed 30 months after the operation have yielded normal findings.

Discussion

Desmoid tumors or musculoaponeurotic fibromatoses are rare. About two-thirds of them occur in the abdominal wall, and they represent only 0.03% of tumors of the abdominal wall. The diagnosis is based on the presence of a painless mass, adherent to the muscle plane, whose size increases with time. Although spontaneous regressions have been described, the current treatment is wide local excision [1–4,6,7].

The etiology of desmoid tumors is unknown, but var-

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Figure 1. *Intraoperative photograph showing the tumor (T) freed from the peritoneal insertion and extending cranially. In the lower portion the distal tubing (D) emerges close to the intestinal folds in the abdominal cavity.*

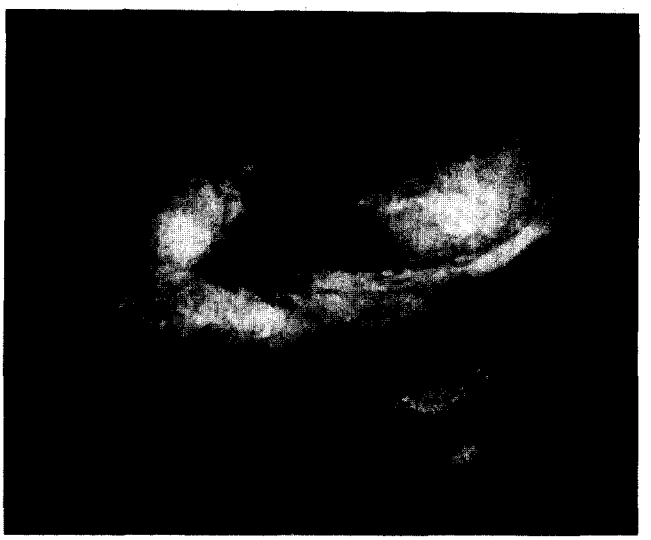
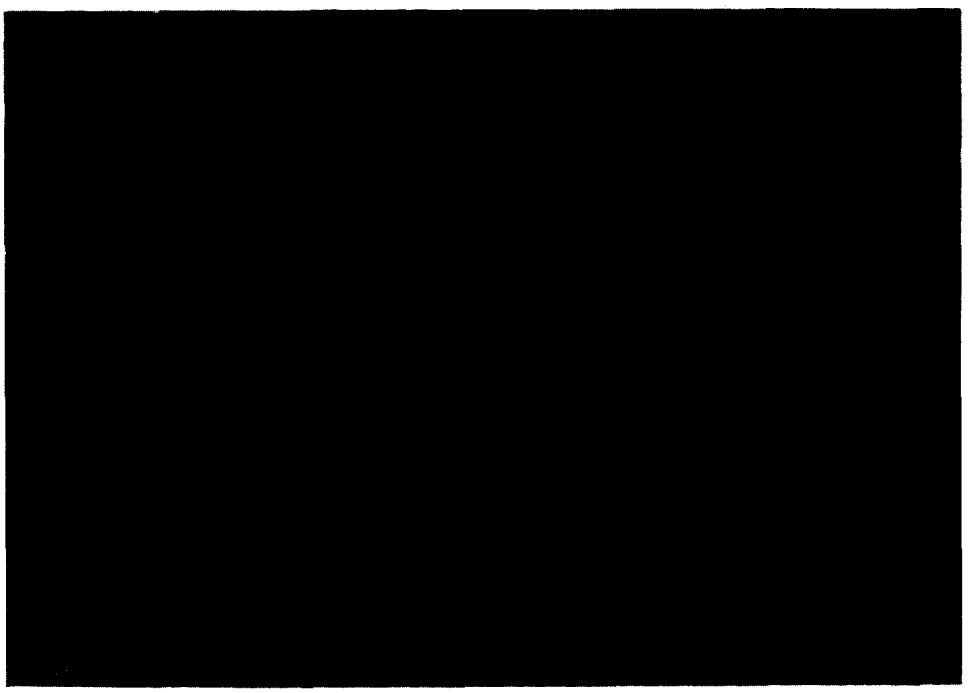


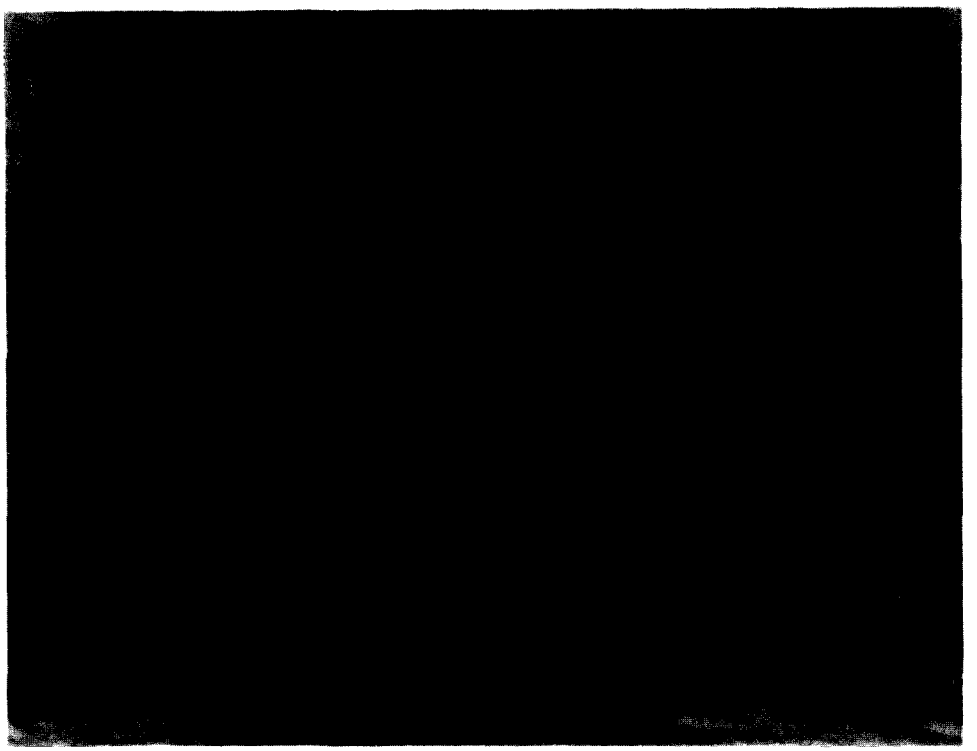
Figure 2. *The removed tumor, with its peritoneal extension below and its cranial extension to the left (bar = 1 cm).*

ious factors (traumatic, endocrine, and familial) are cited; the condition is related to Gardner's syndrome [1–7]. In our patient, the operation performed in order to place the CSF shunt was an evident traumatic occurrence, in addition to the presence of the Silastic distal tubing. The Silastic tubing is not a totally inert material and can produce a fibrous reactive sheath along the tube. In our case, the tumor continued cranially with the fibrous sheath and its implantation in the peritoneum corresponded to the point of entry of the catheter in the abdominal cavity. This suggests the possibility that the tumor arose from and grew in the reactive fibrous tissue surrounding the distal tubing. A similar case has been reported by Jewett and Mead [7], in which a desmoid tumor was found

Figure 3. *Photomicrographs of the desmoid tumor removed. The tumor consisted of fibrous tissue that infiltrated the neighboring muscle (***A***; H&E, ×60), with some mitotic figures (***B***, opposite; H&E, ×120).*



A



B

arising from a fibrous capsule formed around a silicone breast implant.

The mechanism through which the tube became disconnected at the reservoir can be attributed to the stretching of the catheter as it became surrounded, trapped, and displaced by the growing tumor. Then it moved as far as the abdominal cavity, as usually happens.

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