Rupture of spinal dermoid tumors with spread of fatty droplets in the cerebrospinal fluid pathways

Abstract Cranial and spinal MRI was carried out at 0.5 or 1.5 T in five patients with spinal dermoid tumors. Free fatty material was appreciated within the normally communicating cerebrospinal fluid pathways in all five cases and in one case fat droplets were also observed within a dilated central canal of the spinal cord. While dissemination of lipid within the subarachnoid space and ventricles is easily understandable, the presence of lipid droplets within the central canal is more difficult to explain, since the central canal is only potential in the adult. When a dermoid tumor is suspected, we recommend MRI of the entire central nervous system, to detect possible leakage of fat from rupture of a cystic portion of the tumour.

Key words Dermoid tumor, spinal \cdot Syringomyelia \cdot Fat, free \cdot Meningitis \cdot Magnetic resonance imaging

Introduction

Spinal dermoid and epidermoid tumours are rare benign, slow-growing, dysontogenetic tumours arising from inclusion of ectopic embryonic rests of the ectoderm and mesoderm within the spinal canal at the time of neural tube closure between the 3rd and the 5th week of embryonic development [1–3]. It is believed that they may also rarely occur secondary to implantation of dermal fragments into the spinal subarachnoid space by accidental trauma [4], lumbar puncture [5] or surgery [6, 7].

These tumours comprise 0.7–1.8% of intracranial [1, 6, 8–10] and 1–2% of intraspinal tumours [11]. Among dermoid tumours, the spinal location seems to be slightly more common [6, 12] while epidermoid cysts statistically prevail intracranially. There is a slight male predominance, and most dermoid tumours are identified during the 2nd and 3rd decades. Spinal dermoid cysts may be intramedullary, intradural–extramedullary or extradural [12, 13]. They occur predominantly in the lumbosacral region (60%), involving the cauda equina and the conus medullaris, and are quite rare in the upper thoracic (10%) and cervical (5%) regions [12, 14, 15].

Dermoid tumours have the appearance of cysts, typically unilocular, with a creamy consistency, containing yellow or greenish-brown viscous fluid composed of different kinds of lipid (cholesterol crystals, lipid metabolites, keratin). They are surrounded by a wall of connective tissue, thicker than that of epidermoid tumours, lined by stratified squamous epithelium containing dermal appendages such as hair, sebaceous glands, sweat glands and hair follicles and, less commonly, teeth and nails [16]. The high lipid content, derived from the sebaceous secretions, can cause high signal on T1-weighted spin-echo images [8, 11]. The signal may, however, be inhomogeneous, due to the different components within the cyst [17]. Calcification may be sometimes appreciated in its wall, while bone and cartilage may be observed inside the tumour. Dermoid tumours may show two distinct portions: a lipid one and a more solid or more fluid one [12], which may give rise to fluid level.
Dermoid cysts generally grow slowly, and tend to spread in the subarachnoid space. These characteristics explain the relative paucity of symptoms. The spinal mass may grow silently, achieving great size without producing signs of cord or spinal nerve compression.

These tumours often become acutely symptomatic following rupture or infection, which may elicit an inflammatory meningeal reaction, which frequently precedes symptoms and signs due to the space-occupying lesion itself. Leakage of fat into the cerebrospinal fluid (CSF) may also be clinically silent [18]. On the other hand, rupture of the cyst, with dissemination of fat droplets in the CSF pathways may induce nonspecific clinical findings linked to chemical or bacterial meningitis [19–23], headache [24, 25], seizures [26], transient ischaemic attacks associated with arterial spasm [27, 28], or hydrocephalus [29].

There is usually no communication between the cyst and the subarachnoid space; dissemination of fatty material within the subarachnoid space and/or ventricles occurs because of spontaneous, iatrogenic or traumatic rupture [30, 31]. Rupture of intracranial dermoid cysts was once considered a dramatic, fatal event, diagnosed most frequently by necropsy. The first cases of dermoid tumour were reported by Verburus (1745) and Tannehain (1897) [32], each describing two intracranial tumours. A few cases of ruptured dermoid cyst with spillage of lipid into the ventricles were suspected because of a fat-fluid level on conventional skull radiographs [33]. Although MRI has increased incidental detection of asymptomatic spread of fat, there are few reported cases of ruptured spinal dermoid tumours.

Symptoms and signs secondary to the space-occupying lesion are location-dependent and due to the irritative effect on and/or compression of the adjacent structures. Dermoid tumours may be associated with overlying bony malformations, myelomeningocele [23], hydrocephalus and/or a dermal sinus tract. They may be complicated by infection, more frequent in the presence of a dermal sinus, or very rarely by malignant degeneration [34–36].

We report MRI of five patients with spinal dermoid cysts, who underwent MRI examination for various reasons, following spontaneous or iatrogenic rupture of the tumour and dissemination of fatty material.

Case reports

Case 1

A 33-year-old man underwent MRI of the lumbar spine because of progressive paraparesis and sensory disturbances in the legs which began approximately 2 years previously. He complained of back and leg pain, leg weakness and bladder dysfunction. A unilocular intraspinal mass was found at L1, involving the lumbar enlargement, which appeared expanded. The mass gave inhomogeneous high signal on the spin-echo (SE) T1-weighted images, low signal on T2-weighted fast SE (FSE) and gradient-echo images and no contrast enhancement. The lumbar enlargement was isointense with the adjacent spinal cord, apart from linear high signal at its centre, thought to represent oedema or a syringomyelic cavity. Some calcification was detected on axial CT. A large exophytic intramedullary tumour of creamy consistency was excised; histological examination revealed a dermoid cyst. The patient made a good recovery. One year later, MRI showed small foci of high signal within the central portion of the lumbar enlargement of the spinal cord on the SE T1-weighted images (Fig.1). Other small lipid droplets were detected in the central canal up to the level of C6 (Fig.2), in the interpeduncular fossa, interhemispheric fissure and right lateral ventricle. When the patient was placed prone, the intraventricular fat floated up to the occipital horn of the left lateral ventricle.

Case 2

A 61-year-old man had a sudden psycho-organic syndrome, progressive headache and a clinical suspicion of intracranial hypertension. In 1973 he underwent partial resection of a dermoid cyst at the lumbar enlargement; neuroradiological investigation was not extended to other parts of the CNS. In 1993, lumbar MRI, carried out for progressive paraparesis and urinary incontinence, showed a recurrence. The patient underwent radical resection of the dermoid cyst. On the current admission cranial MRI showed a droplet of lipid in the cerebral aqueduct (Fig.3), triventricular hydrocephalus, secondary to stenosis of the aqueduct, and fatty deposits in the interhemispheric fissure, cisterna magna and left lateral ventricle.

Case 3

A 44-year-old man, with a history of hydrocephalus of unknown aetiology (congenital aqueduct stenosis was suspected) treated with ventriculoperitoneal (VP) shunting, was admitted because of sudden sphincter dysfunction and leg weakness. MRI showed a large mass at the lumbar enlargement, made up of two portions: a cranial one isointense with the spinal cord on T1-weighted images, and a caudal one giving high signal on T1 weighting, and low signal on FSE GRE T2-weighted images (Fig.4). The solid portion showed mild, homogeneous contrast enhancement. A small linear area, giving high signal on T2 and low signal on T1 weighting, was seen in the terminal portion of the spinal cord representing a synx and some oedema or pericystic gliosis (Figs.4a, b, 5). Because of our previous experience and suspecting intracranial dissemination of fat droplets as the cause of the hydrocephalus, we performed MRI of the head, which revealed small lipid droplets in the cerebellopontine angle, interpeduncular fossa, cisterna magna, chiasmatic cistern, right lateral ventricle and in the foramina of Luschka, possibly hindering CSF flow at this level (Fig.6). The 4th ventricle appeared enlarged, as did the aqueduct and right lateral ventricle. The VP shunt was in the left lateral ventricle. The intraventricular fat floated upwards on changing the patient’s position from supine to prone (Fig.7). The examination was extended to the cervical and thoracic spine, where fat droplets were detected in the posterior subarachnoid space at T3 and T7–8 (Fig.8). At surgery the lumbar neoplasm was totally removed except for a thin capsule adhering to the lumbar enlargement. Histological examination confirmed a dermoid cyst.