Letters to the Editor

excise the lesions in view of the risks involved. Regular follow ups should ensure that surgical intervention precedes any permanent neurological damage.

Within three months, the patient had fully recovered from his behavioural disturbances. His seizures remained well controlled over the next year. He is still undergoing further follow up.

The medical literature over the past 30 years contains reports of intracranial mucoceles in about 60 patients. Most of the neurosurgical literature involves individual case reports1 and occasional series.2

This particular patient had certain unusual features. Patients in the previous reports presented with associated symptoms of intraorbital or paranasal problems. Intracranial mucoceles presenting with epilepsy3 and frontal lobe signs were exceptionally rare. Also this patient gave no history of orbital, visual, or sinus related symptomatology, previous operations, or trauma.

Although we failed to identify a persistent tract, it is very likely that the mucoceles were associated with the midline fusion defects of bifid uvala, bifid septum, aerated crista galli, and the midline nasal subcutaneous tract. It is probable that trapping of an ectopic focus of functioning respiratory epithelium in the intracranial compartment led to the subsequent production and accumulation of mucus and debris. The high proteinaceous contents would have accounted for the hyperdense images on CT.

The CT appearances satisfy two of the three major criteria suggested by Perugini et al4 for the diagnosis of mucoceles—namely, the presence of a homogenous isodense mass and a clear cut perimeter. The absence of the third criteria of patchy oedema around the mass may have resulted from the distance of the mucoceles from the bony walls. The MRI appearance of mucoceles is more variable5 and in this case did not provide any extra information.

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Irradiation induced osteosarcoma in the posterior cranial fossa six years after surgery and radiation for medulloblastoma

The development of a secondary malignant neoplasm in an irradiated field is a rare but serious complication of therapeutic irradiation. Because of the increased survival rates for patients who receive radiation therapy for malignancies, this problem has become more prevalent.6 Although radiation induced neoplasms can originate virtually from any kind of tissue, the most common are acute myelogenous leukaemia and sarcomas of soft tissue and bone. We report a radiation induced sarcoma of the suboccipital bone after irradiation for a medulloblastoma.

A six year old girl was first seen in 1987 with a history of vomiting, progressive headache, and an unsteadiness during walking. Clinical examination showed slight papilloedema, horizontal nystagmus, and gait ataxia.

Brain CT disclosed a moderate hydrocephalus, caused by a large hyperdense tumour occupying the entire fourth ventricle (fig 1A). The tumour was incompletely removed and histologically classified as a medulloblastoma (fig 2A and B). Radiation therapy of the craniospinal axis with a dose of 34 Gy in daily fractions of 1.8 Gy was given, followed by a surdosage of 20 Gy in 10 fractions at the posterior fossa. During the subsequent years, she was symptom free. Yearly CT showed no recurrence of tumour.

In 1993, at the age of 13, she presented with a two day history of episodes of headache without vomiting or nausea. On the day of admission, she had noticed a painful swelling in her neck.

On examination, a solid lump was palpated at the lower occiput. No neurological abnormalities were found.

Brain CT showed a hydrocephalus caused by a large biconvex, contrast enhanced, and calciferous tumour in the dorsal part of the posterior fossa. The tumour infiltrated the occiput and showed extracranial extension (fig 1B). Magnetic resonance imaging did not show tumour involvement of other parts of the brain (fig 1C) or spinal cord (not shown). The MRI T2 weighted and enhanced images showed a large intracranial extra-axial mass with a heterogeneous appearance (figs 1B and 2B).

The tumour had extended through the foramen magnum and reached the upper cervical spine. The posterior fossa was completely replaced by the mass. The lesion was removed and the patient showed a complete recovery.

The histology revealed the presence of mucus and debris. On electron microscopy, there were epithelial cells with microvilli, indicating the presence of a mucocele.

The patient is doing well at the current follow-up of 16 months.

Histology

Malignant osteogenic sarcoma of the posterior cranial fossa with a mucocele

Two weeks after the operation, the bone window of the CT scan showed a hypodense lucency with a rim of normal bone as a result of radiation induced osteoporosis (fig 1C).

Figure 2 (A) Frontal view showing the midline nasal subcutaneous mucocele. (B) Sagittal CT scan showing the midline nasal subcutaneous mucocele. (C) Axial CT scan showing the midline nasal subcutaneous mucocele.
shown). The ventricles were drained externally and a biopsy of the tumour was taken. Histological examination showed an osteosarcoma. As no long term benefit could be expected from further treatment, the external ventricle drain was removed and the child died soon afterwards.

The tumour removed in 1987 consisted of soft tissue with a greyish aspect. Smear slides were easy to make and showed a highly cellular lesion consisting of cells with round monomorphous nuclei and several mitotic figures (fig 2A). Histology showed an extremely cellular lesion consisting of cells variable in size with scanty cytoplasm. The coarse chromatin of the nuclei was haematoyxphilic. Neuroblastic differentiation manifested by Homer Wright rosettes was absent. Numerous mitotic figures and some vascular and endothelial proliferation were present. Gomori's reticulin and immunohistochemistry for glial fibrillary acidic protein synaptophysin and neurofilament stains were negative. The tumour was diagnosed as a medulloblastoma (fig 2B).

The second tumour, removed in 1993, was completely different. It could not be smeared due to its fibrous, almost hard, structure. Touch preparations did not show the true nature of the tumour. Histology showed the typical appearance of an anaplastic mesenchymal tumour with granular calcifications. Malignant osteoid—that is, poorly formed bone surrounded by "malignant osteoblasts" with hyperchromatic nuclei—was present. Histochemistry showed a strong alkaline phosphatase positivity and immunohistochemistry for S100 showed a reactivity in the chondroid areas. The tumour was classified as an osteosarcoma.

In this child an osteogenic sarcoma was found in a field irradiated six years previously. It was diagnosed as a radiation induced sarcoma because: (1) a sarcoma developed in an irradiated field; (2) a latency period of at least four to five years elapsed from the initial radiation to appearance of the sarcoma; (3) the induced sarcoma was proved histologically to be different from the initial malignancy.

The incidence of radiation induced sarcoma of bone after therapeutic irradiation is low. After five to eight years survival, the risk of developing one varies from 0-05% to 1%.2 Although radiation induced sarcomas, like most spontaneous lesions, do occur in soft tissue, they preferentially originate from bony tissue situated near or in the irradiated field. This can be attributed to the higher absorption of radiation in bony tissue.3 The incidence of postration osteogenic sarcoma steadily increases with the patient’s age until the sixth decade of life, whereas spontaneous sarcomas are most commonly seen in preteen and young teenage patients.4 Spontaneous osteogenic sarcomas are clustered, up to 50%, in the knee region. Flat bones of the pelvis, and shoulder and craniofacial bones are the sites most commonly occupied by postration sarcomas.

Radiation characteristics for the development of radiation induced sarcomas are uncertain. Induction seems to be irrespective of the type of radiotherapy given. Total tumour dose is considered the most important factor. A total tumour dose less than 30 Gy is unlikely to induce radiation induced sarcomas.5 This is based on the finding that sarcomas of bone are usually seen after a full course of radiation, which lies normally between 40–70 Gy.

Tissues undergoing active proliferation are most susceptible to carcinogenic effects. Neoplasms of thyroid and bone are therefore more commonly seen during childhood. In general, children are more sensitive to cancer induction by radiation than adults.6

Within the group of radiation induced sarcomas, osteosarcoma is prevalent in about 50% of the cases, fibrosarcoma in 25%—pleomorphic, spindle cell, chondrosarcoma, and various other types constitute the remainder.

The average latency period for radiation induced sarcomas of bone is 11 years, usually ranging from eight to 16 years.1 According to animal studies, the latency period is inversely related to the radiation dosage.1 A latency period of less than four years makes the diagnosis of radiation induced sarcomas unlikely.

Treatment of postirradiation sarcomas of bone is difficult. Like their spontaneous counterparts, radiation induced sarcomas are radioresistant. Chemotherapy, which can be successful for non-radiation induced sarcomas of bone, is usually ineffective. Because of the poor prognosis of patients with radiation induced sarcomas in bone, in particular of craniofacial bones, most authors advocate more or less radical surgery. Although recommended by some authors, the use of additional radiotherapy or chemotherapy is still a very controversial issue. For our patient, surgical removal could never be radical and no benefit of either radiotherapy or chemotherapy could be expected.

In summary, when pain or swelling occurs in a field irradiated more than four years previously, the presence of a radiation induced sarcoma should be suspected.

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