Primary intraosseous meningioma

J.Sales-Llopis

Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain

Sclerotic tumours of the calvaria are rare and may be due to primary and secondary bone tumors as well as extradural tumours of meningeal origin.

“Primary intraosseous meningioma (PIOM)” is a term used to describe a subset of Primary extradural meningioma that arise in bone, when no dural attachment is present.

They can present either as an osteoblastic lesion or an osteolytic lesion.

Primary intraosseous meningioma on a calvarium from Byzantine Greece is described [1].

231 cases reported in the literature during the CT era.

The constituent ratio of male in head increases markedly and have bimodal distribution of ages. The most common presenting symptom was a mass in the region of the lesion. The average duration of symptom was 2.38 years. The skull convexities, paranasal sinuse and nasal cavity, and middle ear ranked as the top three of all sites of tumors. The most common type was Type II (calvarial or diploic). Among 231 cases, total, subtotal and partial removals of tumors were achieved in 89%, 5.5% and 3.1% respectively, and no death occurred perioperatively in all patients. 90% were benign, 5.6% atypical and 3.9% malignant in the 231 cases. The most common histopathological subtype was meningothelial meningioma. The recurrence and tumor-related death rates were 22.4% and 8.2% respectively during a mean 3.03-year follow-up. The results demonstrate that they have some marked clinical characteristics compared with primary intradural meningiomas. Total tumor removal together with a wide excision of all involved tissues followed by the reconstruction of tissue defects is the best surgical project. The prognoses are good in the benign cases after complete surgical resections [2]

Epidemiology

They account for 2% of all meningiomas [3] , and are generally localized in the frontoparietal and orbital regions.

While most PIMs are located in the sphenoid bone, associated calcifications were visible in 58% of the cases on CT scans [4]

It represents approximately two-thirds of all extradural meningiomas [5]

Both osteolytic radiological features and atypical pathological features are extremely rare

The literature contains no reports clearly detailing how the preoperative diagnosis of primary

intraosseous meningiomas should be made, and they are often mistaken for primary bone tumors and hyperostosis en plaque meningiomas [6) 7) 8) 9) 10)].

Classification

see Primary intraosseous meningioma classification.

see Intraosseous meningioma of the sphenoid bone

Etiology

The origin of primary intraosseous meningiomas remains unknown. Turner et al. have shown the rests of arachnoid cap cells at autopsy [11)] while Azar-Kia et al. proposed that the rests of arachnoid cap cells remained between the sutures during embryogenic development [12) 13) 14)].

The etiology of intraosseous meningiomas that develop in posttraumatic linear fractures can be explained by the same mechanism.

Pathology

Intraosseous calvarial meningiomas are usually easy to diagnose histologically if they are of the meningothelial type; however, they may cause diagnostic challenges when they manifest as unusual morphologic variants, such as the microcystic type.

To address this issue Velázquez Vega et al. present a series of 9 cases of calvarial microcystic meningiomas arising in 7 female and 2 male patients; all patients were adults. The tumors had heterogenous findings on imaging studies and ranged in size from 1.1 to 4.3 cm in greatest dimension. The neoplasms were composed predominantly of stellate and spindle cells with long, thin interconnecting cytoplasmic processes arranged in a complex network. The resulting cellular architecture was “sieve-like” in appearance because of the formation of numerous small “cyst-like” spaces interposed between the cytoplasmic processes of the tumor cells. All of the neoplasms expressed the characteristic immunophenotype of meningiomas (EMA, PR positive). Most tumors were resected, and none of these have recurred during a follow-up period of 1 to 83 months (average 17 mo). The morphology of the tumors and their anatomic location generated problems in diagnosis, especially in 6 patients with a history of malignancy and for whom metastatic disease was suspected clinically. Intraosseous microcystic meningioma is uncommon, and this series, the largest reported to date, describes their clinicopathologic findings, biological behavior, and features that facilitate their accurate diagnosis [15)].

Diagnosis

Primary intraosseous meningioma diagnosis

Differential diagnosis

The differential diagnosis of these lesions is wide and includes not only primary and secondary bone tumours but also extradural tumours of meningeal origin that can involve or arise in skull bones [16) 17) 18)].

The radiological differential diagnosis of osteoblastic lesions should include metabolic diseases including osteoma, hyperparathyroidism, meningioma, A and D hypervitaminosis.

This tumor can be clinically and radiologically confused with metastatic cancer and other malignant bone tumors. Two case report has dealt with 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography/computed tomography (PET/CT) in a primary intraosseous meningioma of the calvarium showing high 18F-FDG uptake [19) 20)]

Intraosseous lipoma [21)].

Treatment

The recommended therapy is surgery, with complete resection whenever possible.

Radiological follow-up is recommended in subtotal removal. Options including adjuvant RT are recommended in recurrent and symptomatic cases.

Outcome

Although most calvarial meningiomas are benign they undergo malignant change more commonly than intracranial meningiomas [22) 23)].

Case reports

A 57-year-old woman presented with an incidentally discovered scalp lump on the head. Neurological deficits were not found. Radiological examination revealed a localized osteolytic lesion in the right parietal bone, which was initially diagnosed as a bone tumor and was surgically resected. At surgery, a tumor mass was found located at the brain convexity without dura attachment. It was tightly attached to the brain parenchyma and had no distinct boundary from the brain. The mass was rather small, but resulted in significant osteolysis of the skull and destruction of the dura. Simpson grade I resection of the tumor was performed. Histological and immunohistochemical results indicated a meningothelial meningioma.

Both preoperative and intraoperative diagnoses are difficult in this case. Knowledge of this case is crucial for clinicians to be aware of this entity because it can be easily confused with bone tumors. Further research on the relationship between meningioma and bone metabolism is required to investigate the mechanism of osteolysis [24)].

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2016

A case of primary intraosseous meningioma (PIM) which arose in the frontal bone of a 63 year old woman who complained of progressive pain and thickening of the right skull. Radiology showed a large osteosclerotic lesion in the right frontal bone. Histology showed an intraosseous lesion containing dense fibrous tissue in which there were scattered cells that expressed epithelial membrane antigen and progesterone receptor. The tumour was partially resected and 3 years after operation has not recurred [25].

2015

Kwon et al. report a primary intraosseous meningioma of a 69-year-old man who had headaches and a mass on right parietal scalp for the past few months. Remarkably, the brain tissue within the osteolytic cavity of the skull was normal in computed tomography and magnetic resonance images. Resection, duraplasty, and cranioplasty were performed. The patient's symptoms disappeared after surgery, and the histological diagnosis was an osseous meningothelial meningioma (World Health Organization grade I) [26].

Bernal-García et al. present the case of a large primary intraosseous osteolytic meningioma within the occipital bone, which was completely excised five years ago, currently presenting no signs of recurrence [27].

A 64-year-old male patient with pain and swelling on the right side of head that had started about 6 months ago and increased slowly. The patient’s neurological and physical examination findings were normal. Radiological examination showed a sclerotic bone lesion causing a spicular periosteal reaction on the right frontotemporal region. As the lesion showed a spicular periosteal reaction, a lesion that does not destroy the bone structure in T1- and T2-weighted images but causes decreased signal and hypointense periosteal reaction in the bone was searched for on contrast cranial MR examination performed to exclude malignancies, and dural thickening and contrast enhancement in this region were searched for on gadolinium T1-weighted images.

These findings were identified as indicative of calvarial metastases or primary osteosarcoma. Three-phase total body bone scintigraphy showed perfusion, and blood pool scans showed increased activity consistent with hyperemia only in the right temporal-frontal region, with increased heterogeneous osteoblastic activity in the same region during the late phase. These findings were identified as indicative of a primary malignant bone lesion. Based on such clinical and radiological findings, the patient was operated with a preliminary diagnosis of calvarial bone metastases. Histopathological

examination showed infiltrative tumoral tissue between cortical bone lamellae in sections sampled after decalcification. The tumor also invaded the peripheral striated muscle tissue. The tumor was made up of monotonous cells with eosinophilic cytoplasm, forming groups and solid islands, with round to oval vesicular nuclei, some of them containing distinct nucleoli with indistinct margins. Tumoral cells immunohistochemically stained with EMA, Vimentin and S-100 (pale). The patient was diagnosed with intradiploic meningioma in light of these data. Dural thickening was identified as indicative of reactional thickening rather than dural involvement. The patient was discharged with no neurological deficits on postoperative day 3. The patient had no complaints at the 2-month clinical follow-up [28].

2014

A 65-year-old woman presented with a 5-month history of a soft mass on the right frontal area. MR imaging revealed a 4 cm sized, multilobulated, strongly-enhancing lesion on the right frontal bone, and CT showed a destructive skull lesion. The mass was adhered tightly to the scalp and dura mater, and it extended to some part of the outer and inner dural layers without brain invasion. The extradural mass and soft tissue mass were totally removed simultaneously and we reconstructed the calvarial defect with artificial bone material. The pathological study revealed an atypical meningioma as World Health Organization grade II. Six months after the operation, brain MR imaging showed that not found recurrence in both cranial and spinal lesion. This report is a case of primary osteolytic intraosseous atypical meningioma with soft tissue and dural invasion [29].

A 44-year-old female patient who had a protruding right eye and headache. MRI showed a large, destructive, heterogeneously well-enhancing soft tissue mass in the right sphenoid bone suggesting malignancy. (18)F-FDG PET/CT showed a hypermetabolic mass in the same site with an SUVmax of 9.1 The pathological diagnosis by surgery revealed that this tumor was a WHO grade I transitional meningioma. This case suggests that primary benign intraosseous meningioma may show high (18)F-FDG uptake mimicking a malignancy [30].

2012

A 68-year-old man presented with a 1-year history of a soft, enlarging mass in the right parietal region. On admission, his neurological examination was within normal ranges. Skull X-rays and a computed tomogram (CT) of the head showed a bony, destructive lesion abutting the scalp and dura in the right parietal bone. Magnetic resonance imaging (MRI) revealed a heterogeneously-enhancing, bony expansile mass in the right parietal bone, extending into intracranial epidural space. Its greatest diameter was about 6 centimeters. The patient underwent surgery for resection of the mass. The scalp and subcutaneous tissues were normal and the outer table of the skull was destroyed by the well-encapsulated, soft mass. The tumor appeared to arise extradurally. Peeling off the outer layer of the dura and inspecting the dura's inner surface did not reveal the tumor. The tumor, including the surrounding normal bone, was totally resected. The permanent section of the specimen showed cellular whorl formation, frequent mitosis (5 mitoses per 10 high-power fields), and necrosis. The biopsy confirmed the tumor as an atypical (WHO grade II) meningioma. The adjuvant treatment was not done because the mass was totally removed. One year later, he complaint palpable mass on previous operation site. Follow-up brain MRI revealed multiple variable sized nodules and masses in whole skull, causing epidural and subgaleal extraosseous mass formations. Biopsy was confirmed as

the same pathology and the patient did not want further treatment for these lesions.

A 74-year-old woman presented with a 5-month history of a soft mass on the left frontal area. Skull X-rays and head CT showed a bony, destructive lesion extending to the scalp and displacing the dura at the left frontal bone. MRI revealed a 4-centimeter, multilobulated, strongly-enhancing mass on the left frontal bone. The mass was located intra- and extra-cranially, with thickening of the adjacent scalp and meninges (Fig. 4B). The tumor had invaded the scalp. The skull was destroyed by the well-encapsulated, soft mass. The mass was adhered tightly at the scalp and dura mater. The lesion was totally removed, including surrounding normal bone. The tumor showed no involvement on the inner surface of the dura. Biopsy showed a papillary (WHO grade III) meningioma with papillary pagrowth pattern and the immunohistochemistry for epithelial membrane antigen was positive. Ki-67 labeling index was 3%. The patient refused adjuvant radiation therapy. Subsequently, at 19- and again at 45-months later, the patient underwent reoperations for recurrent lesions on scalp and skull. Follow-up brain MRI showed a 1 cm, homogenously-enhancing, recurred lesion on her skull, adjacent to the previous lesion. Five years later after the first surgery, the patient experienced back pain that slowly progressed for 4 months. Spine MRI showed an osteolytic lesion on the body and posterior element of the 11th thoracic vertebra). The patient received corpectomy and posterior fusion, with a diagnosis of metastatic papillary meningioma [31)]

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