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CLINICAL CASE

Treatment of adamantinoma of femur with limb preservation. A case report and review of the literature☆

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KEYWORDS
Bone tumour;
Adamantinoma;
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Abstract
Background: Adamantinoma is a rare lesion of low-grade malignancy, and represents 1% of malignant bone tumours of bones, and is mainly located in two regions of the body, jaw (ameloblastoma), and lower extremities. The treatment of choice is surgery due to it being a radio- and chemotherapy-resistant neoplasia.
Clinical case: A 39-year-old male with a history of neonatal hydrocephalus with moderate psychomotor retardation. He began with pain in the posterior region of the left thigh for one year before admission, which was managed as posterior radicular syndrome. He had sudden intense pain on walking that led him to fall over. In the examination, left pelvic limb with deformity in the distal third with increase in volume in the thigh, with pain to palpation, and presence of crackles in the distal third of the femur were observed. A biopsy of the thigh was performed, with subsequent local wide excision + replacement of bone with cadaver bone and a central medullary nail. The final diagnosis was adamantinoma of femur.

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Conclusion: Adamantinomas are rare tumours. It is important to recognise this type of tumour from the beginning, since its prognosis is excellent in initial stages. It is important to have free margins as survival is very high.

PALABRAS CLAVE
Tumor óseo;
Adamantinoma;
Conservación de extremidad

Tratamiento de adamantinoma de fémur con preservación de extremidad. Reporte de un caso y revisión de la literatura

Resumen
Antecedentes: El adamantinoma es una rara lesión de bajo grado de malignidad, representa <1% de los tumores malignos de huesos, se localiza principalmente en 2 regiones del cuerpo, la mandíbula (ameloblastoma) y las extremidades inferiores. El tratamiento de elección es la cirugía ya que se trata de una neoplasia resistente a la radioterapia y quimioterapia.
Caso clínico: Paciente masculino de 39 años de edad con antecedente de hidrocefalia neonatal con retraso psicomotor moderado. Un año previo a su ingreso comenzó con dolor en región posterior del muslo izquierdo, el cual se manejó como síndrome radicular; posteriormente presentó dolor súbito intenso a la deambulación, por lo que tuvo caída de su propia altura. A la exploración de miembro pélvico izquierdo con deformidad en el tercio distal con aumento de volumen en el muslo, dolor a la palpación y presencia de crepitación en tercio distal de fémur. Se le realizó biopsia en muslo, posterior escisión local amplia + colocación de hueso de cadáver con clavo centromedular con reporte definitivo de adamantinoma de fémur.
Conclusión: Los adamantinomas son tumores raros y es importante reconocer este tipo de tumores desde un inicio, ya que su pronóstico es excelente en etapas iniciales. Es importante tener bordes libres ya que la supervivencia es muy alta.

Background

Primary bone tumours are neoplasias that represent less than 0.2% of all malignant tumours. The most common ones are osteosarcoma (35%), chondrosarcoma (30%) and Ewing's sarcoma (16%); adamantinoma only occurs in <1%.[1] The first case reported is attributed to Maier in the year 1900.[2] In 1913, Fisher reported the first tibia adamantinoma.[3]

The origin of this disease is currently unknown. However, there are various hypotheses. Fisher proposed a congenital implantation of epithelial cells[3]; Ryrie,[4] Dockerty and Meyerding[5] are in favour of a traumatic implantation; Lederer and Sinclair,[6] Naji et al.[7] propose a synovial origin. Currently, the theory that states that this disease has an epithelial origin is a subject of debate; during the development of the embryo the basal epithelium shifts, which supports this since the most frequent place is the anterior part of the tibia where there is proximity to the skin.[8] Among the revised cases by Moon and Mori, 60% of 200 cases had a history of trauma in the tumour area.[9]

Clinical case

A 39-year-old male patient with relevant medical history; his mother died of breast cancer. He suffered from neonatal hydrocephalia with moderate psychomotor retardation. The onset of the disease was a year before admission, with pain in the posterior part of his left thigh. It was treated by multiple doctors as radicular syndrome with mild pain improvement. He attended the casualty department due to sudden intense pain during ambulation with no lesion mechanism, causing him to fall from standing height with limb deformity and reduced mobility. During physical examination, a deformity in his left pelvic limb could be observed, located at the distal third of his thigh. There was volume augmentation, pain on palpation on femoral diaphysis and crepitation on the distal third of the femur. Sensitivity was conserved and pulses present were not compromised. Chest X-ray showed no alterations. Anteroposterior and lateral femur X-rays: with cross-sectional breaks in continuity at the distal third of the femur, in pathological area, due to the presence of a radiolucent, medullar, expansive, monostotic lesion of 70 mm × 45 mm. There was periosteal reaction and the transition area was short (Fig. 1).

Magnetic resonance

Medullar lesion in the intersection of the medial third and the distal third of the left femur that was associated with multifragmentary fracture over the pathological area in a neoplastic lesion, without being able to rule out a bone cyst or another pathological process diagnosis (Fig. 2A and B).