Orbital dermoid

B. Denoiseux1, J. Denekens1, F. Van den Bergh2, 4, F.M. Vanhoenacker2, 3, 4

A 40-year-old woman presented with a slowly growing and painless mass located at the superolateral aspect of the right orbit. On clinical examination, the lesion was easily palpable and of firm consistency with smooth borders.

Computed Tomography (CT) showed a small ovoid mass, with a well-defined wall and an intraläsional fat-fluid level (Fig. A, soft tissue window, arrow). The lesion was intimately related to the zygomatico-frontal suture (Fig. B, bone window, asterisk). Magnetic Resonance Imaging (MRI) confirmed the fat-fluid level on the T1-weighted images (WI) (Fig. C, arrow). There were no erosive changes in the adjacent bone, nor any intra-orbital extension.

Based on the combination of characteristic clinical and imaging findings, the diagnosis of orbital dermoid was made.

For cosmetic reasons, the patient was referred for a surgical excision of the lesion.

Comment

A dermoid is a congenital benign neoplasm, derived from ectoderm and mesoderm. It is considered as a choristoma, comprising histologically normal cells in an abnormal location.

Although dermoids may occur in any subcutaneous location, more than 80% are found in the head and neck area, predominantly in the orbital and peri-orbital area. It is one of the most common non-inflammatory and space-occupying orbital lesions in children, accounting for about 2-9% of all orbital masses. However, dermoids are slowly growing and therefore may appear at any age. They develop most commonly adjacent to suture lines, especially at the zygomatico-frontal suture and less frequently adjacent to the nasofrontal suture.

It is believed that during embryogenesis, ectoderm may get entrapped at the site of the suture.

Histologically, the lesion is lined by a keratinized squamous epithelium with intraläsional dermal appendages such as hair follicles, smooth muscle, sweat and sebaceous glands and fibroadipose tissue. The presence of dermal derivate distinguishes dermoids from epidermal cysts. Intralesional accumulation of keratin debris, sebaceous fluid and calcium is typically present in the cavity and can be noted on the imaging.

Superficially located lesions are more frequent than deeply located lesions. On clinical examination, superficial orbital dermoids have well-definable margins, while deep lesions may extend beyond the orbital rim and therefore have incomplete palpable margins. Deep lesions may have dumbbell morphology, with extra- and intra-orbital components. Depending on the size and extension, they can cause ptosis, proptosis and even diplopia. In rare cases the orbital dermoid can rupture, which may cause a marked inflammatory response.

On CT, an orbital dermoid presents usually as a round or ovoid mass at the superolateral orbital rim near the zygomatico-frontal suture. The majority of the lesions have a well-defined wall. The contents of the lesion depends on the amount of lipid and keratin within it. Intraläsional fat or a fat-fluid level may enhance the specificity of the diagnosis. Calcifications are rare. CT is the preferred technique to demonstrate the intimate relationship with adjacent sutures and any associated erosive bone changes.

MRI may confirm the T1-hyperintense fat component and the fat-fluid levels. Hypointense components may be due to intraläsional keratin debris or calcifications. After administration of gadolinium contrast, there is subtle rim enhancement of the lesion wall but lack of any central enhancement.

It is important to determine whether the cyst is superficially or deeply located, because of the different therapeutic approach. In this regard, imaging plays a pivotal role for precise assessment of intra-orbital or even intracranial extension. Although superficially located asymptomatic lesions can be left alone, they often cause cosmetic problems and therefore surgical excision is preferred. Deep lesions require a more complex orbital dissection and recurrence rate is considerably higher due to partial excision of the lesion wall.

Reference