



Nasoethmoidal Dermoid Cyst With Intracranial Extension. Case Presentation and Surgical Management

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Abstract: Nasoethmoidal dermoid cysts are rare congenital ectodermal malformations. Unlike other dermoid cysts, those located in the midline can exhibit intracranial extension. Clinically, they may manifest with sebaceous discharge through a fistulous orifice and other morphologic alterations, occasionally presenting with complications at the onset. The authors present a clinical case of a nasoethmoidal dermoid cyst with an intracranial extension, treated with combined interdisciplinary surgical intervention. An adequate imaging evaluation before the surgical planning and complete lesion resection are crucial, considering their high recurrence rates.

Key Words: Intracranial extension, nasal fistula, nasoethmoidal dermoid cyst

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Nasoethmoidal dermoid cysts are rare congenital ectodermal malformations, which predominantly occur in males. Nasal dermoid cysts constitute 13% of all dermoid cysts, which typically present as midline masses, particularly along the nasal dorsum, extending from the glabella down the nasal bridge to the base of the columella. Lesions at the nasal tip are considered uncommon in nasal dermoid cysts.¹ They can present as sinus tracts or fistulas, often with or without a noticeable swelling. These lesions are characterized by a fibrous capsule, well-defined borders, lined with squamous epithelium and containing a yellowish pasty material, often interspersed with hair, sebaceous glands, and sweat glands.²

In contrast to dermoid cysts in other locations, nasoethmoidal dermoid cysts can exhibit intracranial extension, as reported in up to 10% of the cases.³

Hence, it is imperative to conduct a thorough imaging assessment to rule out any intracranial involvement before the surgical planning. A complete lesion resection is strongly recommended, given the high recurrence rates documented to date.^{4,5}

In the following sections we present a clinical case of a nasoethmoidal dermoid cyst with an intracranial extension. We discuss the diagnostic process, surgical treatment and the patient's presenting symptoms, providing an updated overview to the information available in the literature.

CLINICAL CASE

A female patient, 14 years old at the time of the visit, presented with aesthetic discomfort related to her nasal morphology without any associated symptoms. Physical examination revealed mild hypertelorism, a prominent forehead, a thick nose with a high nodular dorsum and a broad nasal pyramid. An orifice in the skin was observed in the transition zone between the nasal tip and columella, with the presence of a thick sebaceous-type yellowish secretion upon digital pressure. A computed tomography (CT) scan of the paranasal cavities and a magnetic resonance imaging (MRI) of the face were requested due to the lesion being localized in the midline of the face. The imaging assessment reported the presence of a cystic lesion extending caudally, almost reaching the epidermal plane of the nasal tip with a distal sinus tract at the level of the nasal tip and columella. It passes beneath the nasal bone, leading to the involvement of both nasal bones and its cartilage septum, a descend of brain tissue of the right rectus gyurus through the widened foramen coecum was also observed as shown in Supplemental Figure 1, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>. Both studies suggested a dermoid cyst with intracranial extension. Interdisciplinary surgical management of the lesion was planned, involving teams from Plastic Surgery, Neurosurgery and Otorhinolaryngology.

Supplemental Figure 1, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>. (A) Axial section of face MRI showing a bifid nasal septum and a bony diastasis of the nasal pyramid secondary to the cystic lesion. (B) Sagittal section of the MRI demonstrating a nasal cystic mass with a fistulous tract originating from the dermis of the nasal tip with an intracranial extension.

Surgical Technique

A combined surgical approach was planned, consisting of a bicoronal incision with a frontal craniotomy for the transcranial removal of the lesion, and a direct nasal approach involving an open rhinoseptoplasty. The cutaneous orifice was isolated separately from the skin flap.

A short fistulous tract was dissected in the subcutaneous tissue, which continued into a large irregular cystic mass ex-

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tending from the nasal tip to the glabellar area. The alar complex was luxated and displaced laterally. In the mid-nasal area, a bifid nasal septum was observed due to the mass effect of the cyst along with disruption and diastasis of the nasal cartilage and bone as shown in Supplemental Figure 2, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>.

Through the nasal approach, a complete excision of the cyst and the adjacent capsule was achieved, extending up to the nasal glabellar level.

Supplemental Figure 2, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>: (A) Preoperative image of the cutaneous orifice in the columella-nasal tip transition zone (arrow). (B) Intraoperative nasal image showing the dermoid cyst and its contents adhering to the skin flap (arrow). (C) Intraoperative nasal image showing the displaced lower alar complex, bifid septum, and bony diastasis of the nasal pyramid due to the mass effect of the cyst.

The bicoronal approach provided access to the frontal bone, allowing for a frontal craniotomy through a bony window and removing the bone segment affected by the lesion, including the foramen coecum as shown in Supplemental Figure 3, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>.

Supplemental Figure 3, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>. (A, B) Frontonasal area exposed through the bicoronal approach, highlighting the destructive lesion of the nasal skeleton (arrow) and its intracranial extension. (C) View of the inner side of the excised frontal bone segment with a bony impression due to the cystic lesion.

After completing both combined approaches, a complete excision of the lesion and its capsule was achieved.

Subsequently, the nasal reconstruction phase began. Reduction of the width of the nasal pyramid was achieved, through an osteotomy of the nasal bone with a direct visualization of the upper third of the nose, utilizing the coronal access. As for the lower two-thirds of the nose, through a conventional rhinoplasty approach. The cartilaginous nasal dorsum was reconstructed by joining both sheets of the bifid septum and the upper lateral cartilages with non-absorbable sutures (nylon 5-0). Finally, the complex of lower alar cartilages was reconstructed by connecting both medial crura and domes with nylon 5-0 sutures to achieve a proper nasal tip projection. The nose was fixated with an external nasal splint for 10 days.

The histopathologic examination of the sample confirmed the diagnosis of a dermoid cyst.

The patient had a favorable outcome with satisfactory aesthetic and functional results, without any postoperative complications or recurrences. A clinical and radiologic follow-up for a period of 10 years demonstrated adequate frontonasal bone remodeling as shown in Supplemental Figure 4, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>.

Supplemental Figure 4, Supplemental Digital Content 1, <http://links.lww.com/SCS/H22>. (A, B) Preoperative frontal and lateral images of the nasal area showing a thick nasal pyramid and a nodular dorsum. (C, D) Postoperative frontal and lateral nasal images showing reduced nasal width and a harmonious dorsum without increased volume.

DISCUSSION

Nasoethmoidal dermoid cyst is a congenital midline malformation that can present as a cyst, sinus or a fistula, which in some cases may have an intracranial extension.

Most nasoethmoidal dermoid cysts are diagnosed within the first 3 years of life, as observed in our previous experience.² However, there are cases in which they are diagnosed in adulthood.^{2,4}

Our previous experience, limited to localized nasoethmoidal dermoid cyst,² described an incidence of up to 45%. However, recent publications have shown in a larger case series an incidence of up to 10%.⁴

The pathogenesis is explained by incomplete obliteration of the neuroectoderm during the development of the frontonasal process between the fourth and eighth weeks of gestation. This remnant is covered by a stratified squamous epithelium containing ectodermal and mesodermal elements, such as sebaceous glands.

Clinically, nasal dermoid cyst usually manifests as a subcutaneous mass along the nasal dorsum, with the nasal tip being an infrequent location, which rarely occurs on the nasal tip and nasal septum at the same time.^{1,6} Nasal dermoid cyst causes a deformation of the nasal skin and is rarely asymptomatic, it may manifest with intermittent discharge of sebaceous or purulent material, osteomyelitis of underlying bony structure, or as already mentioned with a deformity, like widening the root or the nasal bridge. It could also cause local or intracranial infectious complications.^{2,3,5,6}

In the case series reported by Hartley et al in 2014, which had a population of 103 patients diagnosed with a nasal dermoid cyst, the authors highlighted the nasal dorsum being the most common location, which was observed in 78 patients, followed by the glabella in 13 patients. Less frequent presentations were found in the columella and the nasal tip, which was observed in only 2 patients in this case series.

In addition, this case series reported an intracranial extension in 10% of the cases (8% extradural and 2% intradural). There are few reports in the literature that mention its clinical manifestation with sinus tracts in the nasal tip or columella as in this case.³

Therefore, the presentation of the nasal dermoid cyst in our clinical case is considered to be rare as shown in the literature mentioned above, considering its topographic location and having an intracranial extension. The dermal location and the clinical signs are not always indicative of the degree of tissue penetration,⁷ they can involve deep structures, including brain tissue, due to the intracranial extension, as demonstrated in our case. The largest case series reported up to 10% of nasoethmoidal dermoid cysts with an intracranial involvement.³ This type of presentation can predispose patients to local infectious complications or brain-related complications such as meningitis and brain abscess.^{3,5} The eradication of these malformations could be considered a preventive behavior for intracranial infectious complications.

For this reason, nasoethmoidal dermoid cysts require an adequate imaging evaluation to determine their location and extent, to assess neighboring structures and to rule out any associated malformations, which can occur in up to 19% of the cases, as reported in our previous experience. High-resolution CT and MRI scans are recommended for the surgical planning to enable a complete lesion removal.²

In the differential diagnosis of midline nasal lesions, consideration should be given to epidermoid cysts, gliomas, meningoencephalocele, tumors of vascular origin, fibromas, neurofibromas, osteomas and lipomas.²

When dealing with a dermoid cyst with an intracranial extension, a combined approach, both bicoronal and nasal, is to be considered, as recommended in the literature. Hartley et al described for these cases a frontal or frontonasal craniotomy, typically combined with an external nasal approach to the cyst.³ Rahbar et al⁸ proposed a treatment diagram of the nasal dermoid, recommending a combined intracranial-extracranial approach in cases, where intracranial extension is observed on preoperative MRI. His surgical approach complies with the approach we applied in our patient.

Pfaff and colleagues presented in 2013 a series of 3 clinical cases in pediatric patients with nasoethmoidal dermoid cyst with an intracranial extension, describing the surgical approaches used to access these lesions. The used approaches were isolated open rhinoplasty access, isolated bicoronal access, and a combined bicoronal and rhinoplasty approach, which were considered the main treatment approaches. This article also reviews the literature on the approaches reported by various authors, mentioning accesses such as rhinoplasty, bifrontal craniotomy (bicoronal incision), frontonasal osteotomy, transglabellar/subcranial osteotomy, anterior small window osteotomy, and transnasal endoscopic, along with modifications or combinations of those approaches.⁹

Considering the location and extension of the nasoethmoidal cyst in our study, a combined approach was performed, as described in the literature, which considered to be a reliable approach to achieve complete lesion removal.

In 2020 Naina and colleagues conducted a retrospective 10-year study, diagnosing 25 children with nasal dermoid cysts, with an average age of 3.7 years. Of these, 4 patients were presented with intracranial extension. They described that the chosen surgical technique was based on factors such as the location and extent of the lesion, radiographic extension of the sinus tract, and the presence of an external cutaneous pit/cyst. In all described cases similar to the one we reported, a combined transcranial and transnasal approach was suggested, with a bicoronal incision and a midline vertical nasal incision.¹⁰

Complete excision of the nasal dermoid lesion, regardless of its extent, is essential to prevent recurrence and infectious complications of the structures involved in its path, including the brain and its membranes. Incomplete removal can result in up to 50% to 100% recurrence of the lesion.^{3,11} There are no reports in the literature regarding the recurrence of dermoid cysts with intracranial extension; our experience with only one case showed a favorable outcome without any recurrences or complications to date.

CONCLUSION

Surgical management with a combined approach and interdisciplinary focus is the recommended method for a complete

removal of a nasoethmoidal dermoid cyst with intracranial extension. We suggest multidisciplinary management including teams from neurosurgery, otolaryngology and plastic surgery.

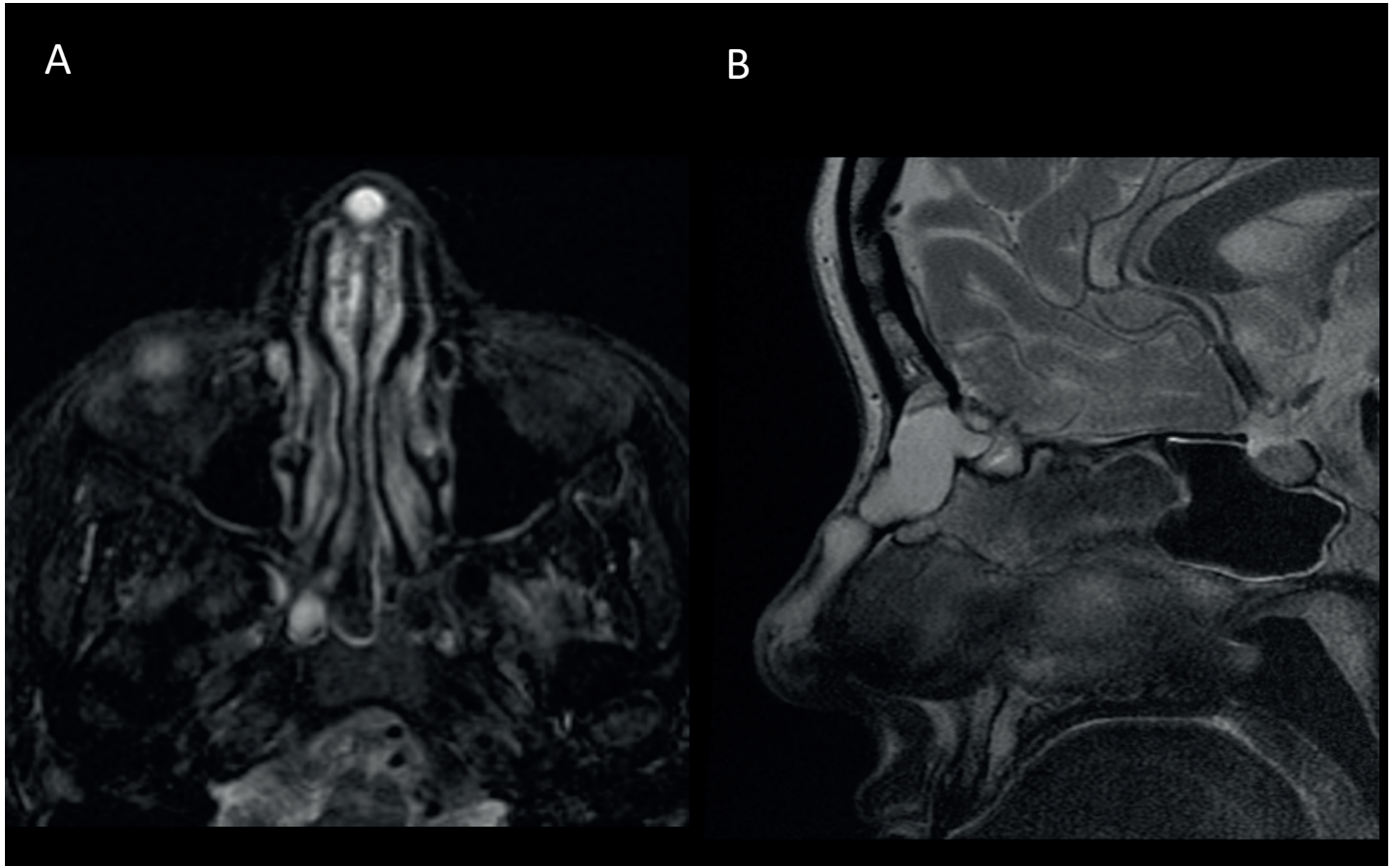
In cases of nasoethmoidal dermoid cysts or any congenital anomalies in the midline facial area, a CT and MRI scans must be performed, to rule out the intracranial extension.

Long-term follow-up is important to evaluate the clinical and radiologic signs of potential recurrences.

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Figura 1



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Figura 2

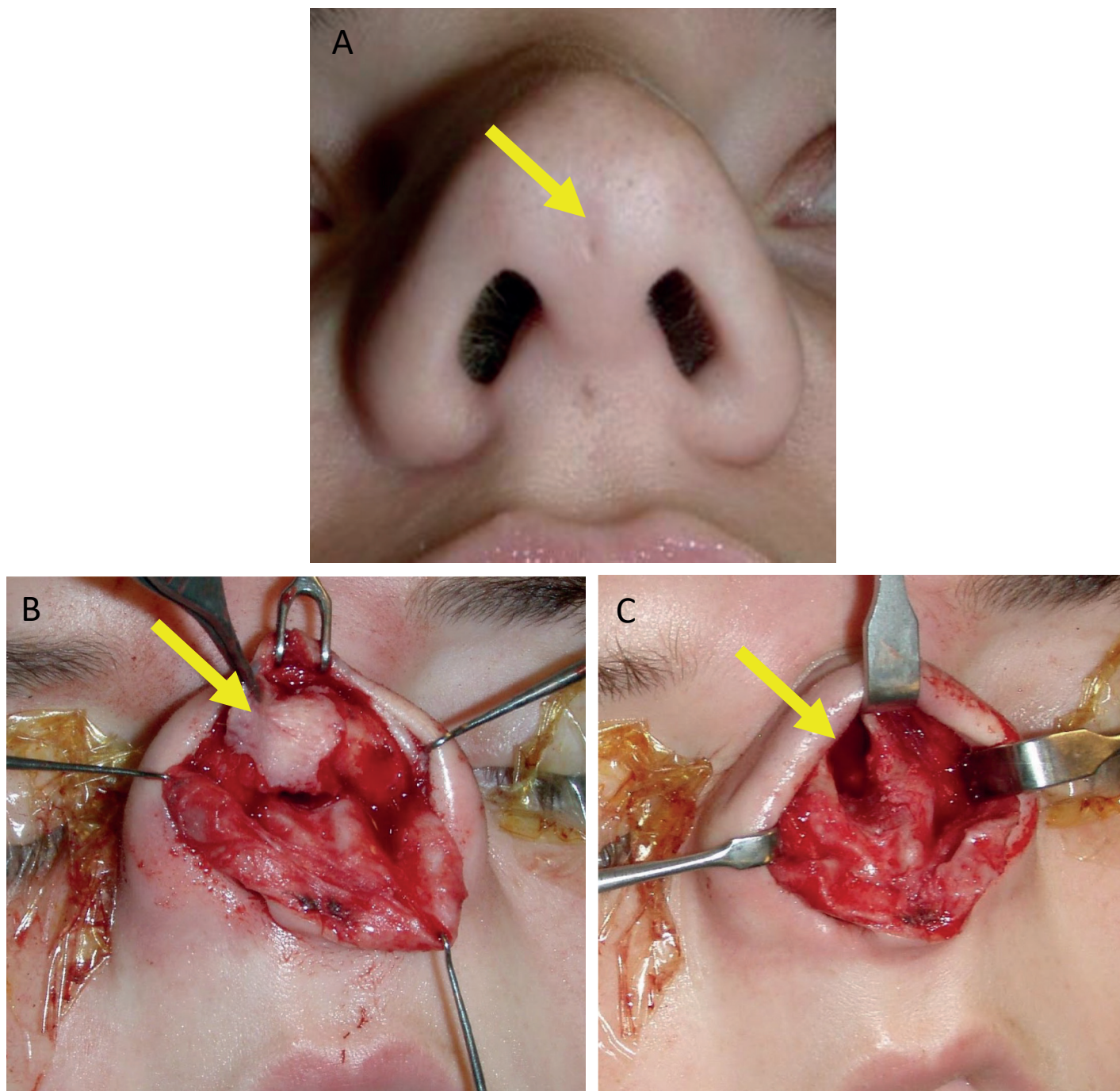
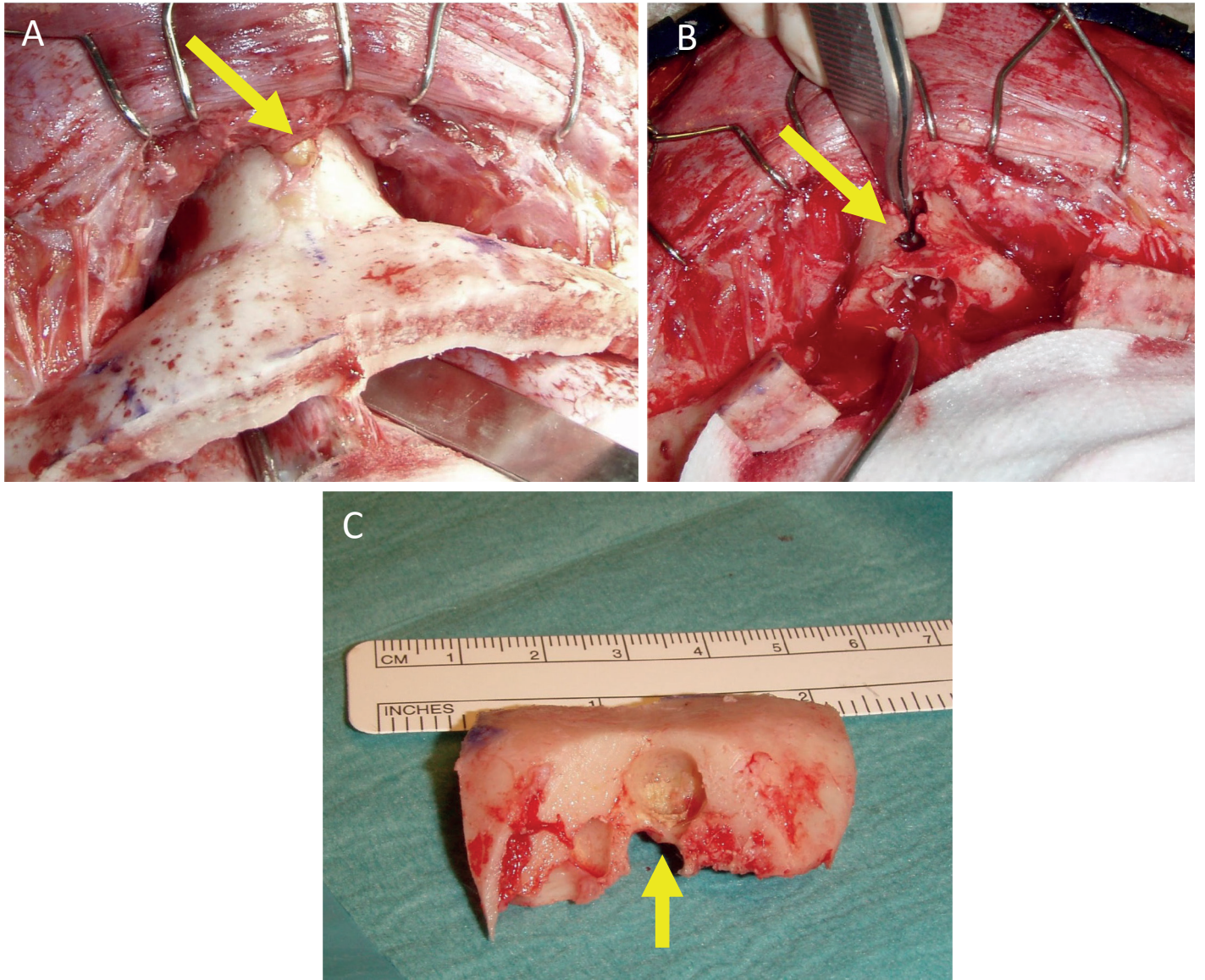


Figura 3



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Figura 4

