

Clinical Report

Epineurial Pseudocyst of the Intratemporal Facial Nerve: A Case Series Study

Stefan Delrue , Tom Cammaert , Philippe Heylbroeck , Marc Lemmerling 

Department of Otorhinolaryngology, Head and Neck Surgery, Sint-Lucas Hospital, Ghent, Belgium (SD, TC, PH)

Department of Radiology, Sint-Lucas Hospital, Ghent, Belgium (ML)

Department of Diagnostic Sciences, Faculty of Medicine and Health Sciences, Ghent University, Ghent, Belgium (ML)

ORCID iDs of the authors: S.D. 0000-0002-1215-060X; T.C. 0000-0001-8975-7357; P.H. 0000-0003-1528-4336; M.L. 0000-0002-6066-1025.

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OBJECTIVES: The goal of this case series was to describe the clinical and radiological characteristics of epineurial pseudocysts of the intratemporal facial nerve (EPIFs) and to discuss the relevance in clinical practice.**MATERIALS and METHODS:** A retrospective case series of 10 consecutive patients with EPIFs identified through computed tomography (CT), between 2009 and 2018. Morphological characteristics, coexisting pathology, facial nerve function, and evolution over time were analyzed.**RESULTS:** A unilateral EPIF was found in 5 patients (50%) and a bilateral EPIF was found in the other 5 (50%). The largest dimensions were observed in the coronal plane, with an average craniocaudal length of 6.0 mm (range, 3–9 mm). None of the patients presented with facial nerve dysfunction. Growth could not be observed in any of the patients. In 5 cases (33.3%), CT imaging showed a reduced transmastoid access to the facial recess caused by the EPIF.**CONCLUSION:** All EPIFs in this study were incidental findings. Facial nerve function was normal in all patients. Knowledge of EPIFs is important to perform safe cholesteatoma and cochlear implant surgery and to prevent unnecessary follow-up imaging.**KEYWORDS:** Facial nerve, mastoid, tomography, X-ray computed, magnetic resonance imaging

INTRODUCTION

An epineurial pseudocyst of the intratemporal facial nerve (EPIF) is a rare lesion of unknown embryologic origin, located adjacent to the mastoid segment of the facial nerve. The term EPIF was coined by Pertzborn et al.^[1] in a case series of 7 cases in 2003. Histopathologic examination showed that EPIFs consist of dense fibroadipose tissue without a true cystic wall. The embryologic origin of these pseudocysts remains unknown^[1]. On temporal bone computed tomography (CT) imaging, an EPIF may mimic an opacified mastoid air cell, a cholesteatoma, or a tumor^[2]. However, the typical location alongside the mastoid portion of the facial nerve, the eccentric morphology, and the small overlying layer of bone allows to differentiate EPIFs from other lesions^[1,3].

MATERIALS AND METHODS

A retrospective radiographic review of all patients diagnosed with an EPIF on multislice CT (MSCT) or cone beam CT was performed between 2009 and 2018. The general policy of the department to perform temporal bone CT imaging was the presence of an air–bone gap (ABG) of at least 15 dB at multiple frequencies and normal otomicroscopy. Other indications were chronic middle ear disease or cases in which inner ear disorder was suspected. All scans were re-evaluated by an experienced head and neck radiologist (M.L.) with a special interest in temporal bone imaging. Only nodular and sharply delineated cystic lesions, enveloped by a thin layer of bone, located in a well-aerated temporal bone adjacent to the mastoid part of the facial nerve were included in the study. Characteristics on CT and magnetic resonance (MR) and the impact on facial nerve function were described. Air conduction (AC), bone conduction, and ABG at the time of diagnosis were calculated for the 0.5, 1, 2 and 4 kHz hearing thresholds according to the guidelines of the Committee on Hearing and Equilibrium.^[4] All procedures performed in this study were in accordance with the ethical standards of the institutional committee and with the 1964 Helsinki declaration and its later amendments.

Corresponding Address: Stefan Delrue E-mail: stefan.delrue@azstlucas.be

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RESULTS

Between 2009 and 2018, an EPIF was identified in 10 patients. In 5 of these patients (50%), the EPIF was found bilaterally. The male to female ratio was 1:1 and the mean age was 35.7 years (range, 9–65 years). In all patients, CT imaging had been performed to evaluate hearing loss. The mean AC was 29.0 dB HL (range, 7.5–72.5 dB HL) and the mean ABG was 6.5 dB (range, 0–17.5 dB). Bilateral fenestral otosclerosis was found in 2 patients (20%). Mean clinical follow-up of patients was 29 months (range, 1–97 months). Facial nerve dysfunction was not observed and there was no indication for a surgical exploration in any of the patients. The average dimensions (anteroposterior × laterolateral in the axial plane × craniocaudal in the coronal plane) of the EPIFs were 4.3 mm (range, 3–7 mm) × 4.4 (range, 2–8 mm) × 6.0 mm (range, 3–9 mm), respectively (n=15). Additional MR imaging was available in 4 patients (40%). These MR scans were performed for neurological reasons not related to the EPIF. In one patient, this showed signs of chronic brain ischemia. Apart from the EPIFs, all other scans showed no abnormalities. All EPIFs were hyperintense on T2-weighting. On T1-weighting, 2 had a hyperintense appearance and 4 a mixed isointense/hyperintense appearance. Intravenous gadolinium was administered in 2 of the patients (3 cases). This could not show enhancement of the lesions. Of the 15 EPIFs, 6 had a posteromedial position, 4 had a posterior position, 3 had a posterolateral position, and 2 had a lateral position relative to the mastoid segment of the facial nerve canal. In 5 ears (33.3%), CT imaging suggested a partial or complete blockage of the transmastoid access to the facial recess. These were the cases with the posterolateral and lateral location. In 3 patients (5 cases), a second CT scan was available, with an interval of 4, 6, and 7 years, respectively. These additional CT scans showed exactly the same lesions and did not indicate any growth. An overview of the patient characteristics is shown in Table 1.

DISCUSSION

EPIFs were first described by Pertzborn et al. in 2003 and subsequently by other authors^[1–3,5]. Although considered to be rare, these lesions may be easily overlooked on CT imaging performed for other reasons. Diseases that may appear similar on CT are mucocoeles, cholesteatomas, neoplasms (hemangiomas, schwannomas, parotid malignancy with perineural spread), and neuromas. The specific location adjacent to the mastoid portion of the facial nerve, the nodular and well-delineated shape, the thin overlying layer of bone, and the stable appearance over time are key features to distinguish EPIFs from other pathologies (Figure 1). Whereas traumatic or inflammatory neuromas are the result of prolonged contact with an inflammatory process, EPIFs are mostly found in well-aerated temporal bones.

MAIN POINTS

- Epineurial pseudocysts of the intratemporal facial nerve (EPIFs) are unilateral or bilateral lesions of unknown embryologic origin.
- EPIFs mostly are accidental findings and remain stable over time.
- Knowledge of these lesions is important to secure safe mastoid surgery and to prevent unnecessary follow-up.

Facial nerve hemangiomas are normally located in the geniculate ganglion area or internal auditory canal and present with facial nerve paresis early in their natural history^[6]. Facial nerve schwannomas have a tubular shape and are often found along multiple segments of the facial nerve. In the mastoid, they can have irregular margins when they break into surrounding cells. They often present with facial nerve symptoms^[7]. Parotid malignancies with perineural spread are irregular lesions extending through the stylomastoid foramen. They typically present with progressive facial nerve paresis and have a primary lesion in the parotid gland^[8]. On CT imaging, it is important not to confuse the stapedius muscle with an EPIF. The stapedius muscle always lies in a medial position relative to the mastoid segment of the fallopian canal, whereas EPIFs are never located medial to the

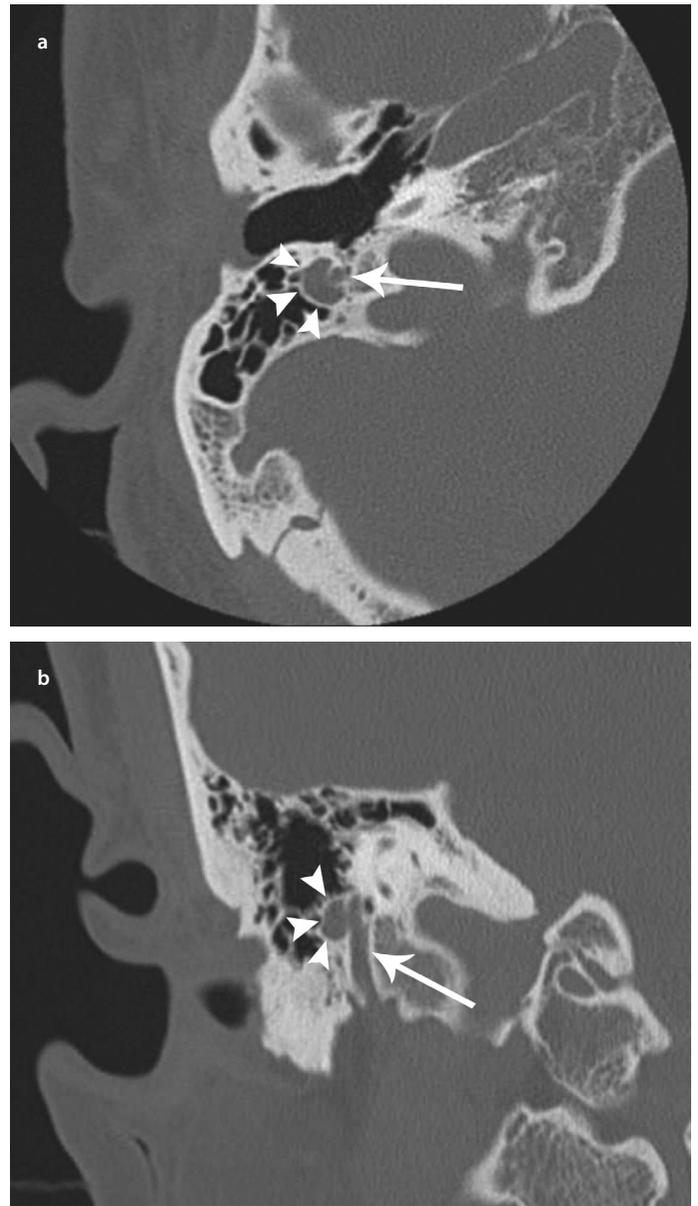


Figure 1. a, b. a) This axial MSCT image of the right temporal bone performed in a 42-year-old male patient shows an EPIF (arrowheads) adjacent to the mastoid portion of the facial nerve (arrow). b) On this coronal MSCT image of the same patient the EPIF (arrowheads) can also clearly be identified lateral to the facial nerve canal (arrow).

canal. Moreover, the stapedius muscle can be followed to within the pyramidal eminence.

In our study, all EPIFs were hyperintense on T2-weighted MR imaging, whereas on T1, they had a hyperintense or isointense/hyperintense appearance (Figure 2). The latter finding probably reflects the fibroadipose nature of such pseudocysts: the T1 hyperintense parts correspond with the fatty components, and the isointense parts with the more fibrous components. There was no contrast enhancement in the 2 patients (3 cases) with gadolinium administration.

It is still unknown whether EPIFs may affect facial nerve function or not. In our series, none of the patients had facial nerve problems, whereas in the original article by Pertzborn et al.^[1], 3 of 7 patients had

facial nerve symptoms (mild spasticity, recurrent facial palsy, and facial pain). However, the latter authors concluded that the EPIFs were probably not the cause because historically, they have never been reported during facial nerve explorations performed for spasticity or recurrent facial palsy.

The embryologic origin of EPIFs is still unknown. Histopathologic studies by Pertzborn et al. showed that they consist of a dense fibroadipose tissue without a true cystic wall.^[1] In our study, half of the patients had a bilateral EPIF. This bilateral occurrence had been described earlier and is an additional characteristic to distinguish EPIFs from other pathology (after exclusion of neurofibromatosis type 2)^[3, 5]. How to explain the bilateral occurrence embryologically remains unclear.

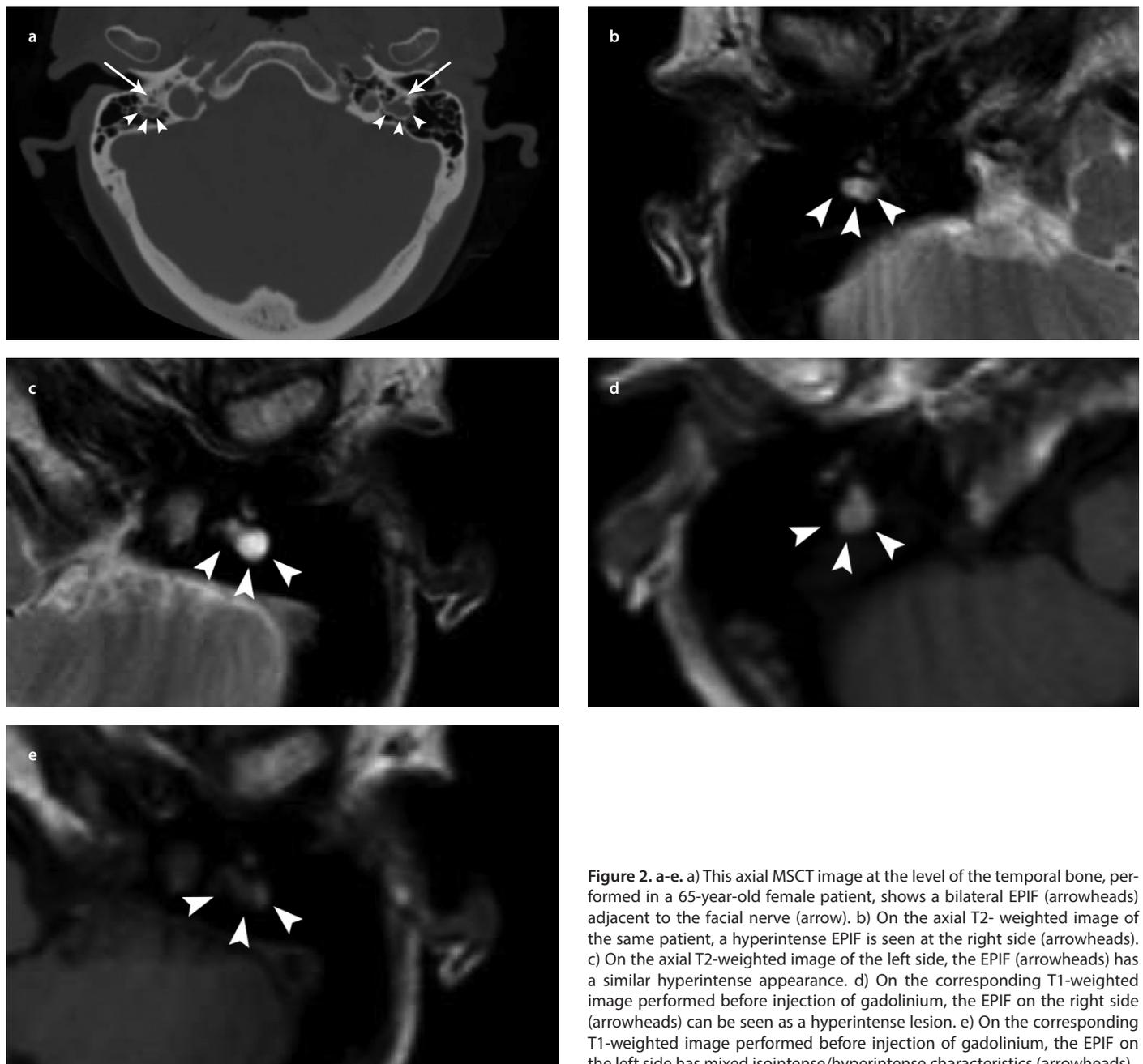


Figure 2. a-e. a) This axial MSCT image at the level of the temporal bone, performed in a 65-year-old female patient, shows a bilateral EPIF (arrowheads) adjacent to the facial nerve (arrow). b) On the axial T2-weighted image of the same patient, a hyperintense EPIF is seen at the right side (arrowheads). c) On the axial T2-weighted image of the left side, the EPIF (arrowheads) has a similar hyperintense appearance. d) On the corresponding T1-weighted image performed before injection of gadolinium, the EPIF on the right side (arrowheads) can be seen as a hyperintense lesion. e) On the corresponding T1-weighted image performed before injection of gadolinium, the EPIF on the left side has mixed isointense/hyperintense characteristics (arrowheads).

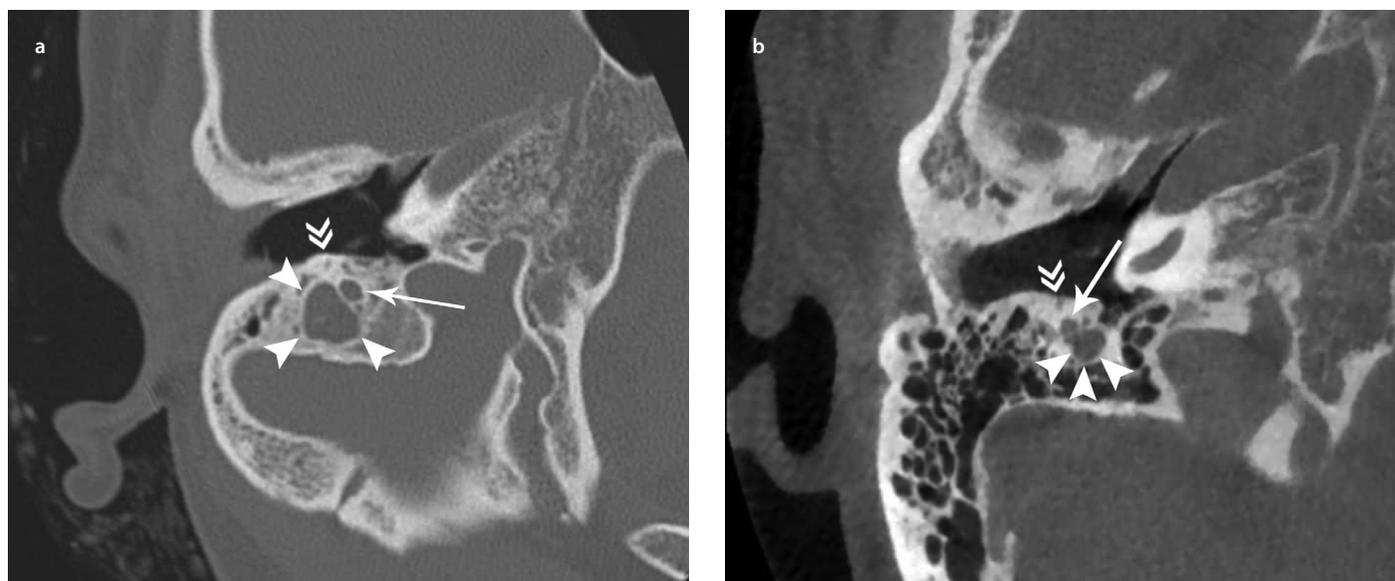


Figure 3. a, b. a) This axial MSCT image performed of the right temporal bone in a 33-year-old female patient shows a large EPIF (arrowheads) posterolateral to the facial nerve canal (arrow), and posterior to the chorda tympani (double arrowhead). Transmastoid access to the facial recess is reduced by the lesion. b) This axial MSCT image performed of the right temporal bone in a 35-year-old male patient shows a smaller EPIF (arrowheads) posteromedial to the facial nerve canal (arrow), not hampering access to the facial recess. The chorda tympani (double arrowheads) lies relatively far from this lesion.

Table 1. Characteristics of patients with EPIF

| Patient | Age* | Sex | Side | Dimensions AP ^a , LL ^a , CC ^b (mm) | MRI | Relation to facial nerve canal | Access to facial recess | Other pathology |
|---------|------|-----|-------|------------------------------------------------------------------------------------|------------------------------------------------------------------|--------------------------------------|----------------------------|--------------------|
| 1 | 33 | F | R | 7, 6, 5 | No | Posterolateral | Reduced | No |
| 2 | 35 | M | L + R | L (2009): 5, 4, NA R (2009): 3, 3, 5 L (2013): 5, 4, 6 R (2013): 3, 3, 5 | No | L: posteromedial R: posteromedial | L: normal R: normal | BFO |
| 3 | 42 | M | R | 5, 3, 5 | No | Lateral | Reduced | No |
| 4 | 30 | M | L + R | L: 3, 2, 4 R: 4, 4, 7 | No | L: posteromedial R: posteromedial | L: normal R: normal | BFO |
| 5 | 65 | F | L + R | L: 5, 8, 6 R: 4, 5, 6 | L: T2 hyper, T1 iso/hyper R: T2 hyper, T1 hyper | L: posterior R: posterior | L: normal R: normal | No |
| 6 | 58 | M | L | 5, 3, 7 | T2: hyper, T1: hyper, NGE | Posteromedial | Normal | No |
| 7 | 34 | F | L + R | L (2010): 5, 5, NA R (2010): 4, 6, NA L (2017): 5, 5, 9 R (2017): 4, 6, 5 | L: T2 hyper, T1 iso/hyper, NGE R: T2 hyper, T1 iso/hyper, NGE | L: posteromedial R: posterior | L: normal R: normal | No |
| 8 | 22 | F | L + R | L: 4, 3, 6 R: 3, 2, 3 | No | L: lateral R: posterolateral | L: reduced R: reduced | No |
| 9 | 9 | F | R | 4, 6, 8 | No | Posterolateral | Reduced | No |
| 10 | 29 | M | R | (2010) 4, 6, 9 (2016) 4, 6, 9 | T2 hyper, T1 iso/hyper | Posterior | Normal | No |

AP: anteroposterior; BFO: bilateral fenestral otosclerosis; CC: craniocaudal; EPIF: epineurial pseudocyst of the intratemporal facial nerve; F: female; L: left; LL: laterolateral; M: male; MRI: magnetic resonance imaging; NA: not available; NGE: no gadolinium enhancement; R: right.

^aAxial plane

^bCoronal plane

*at time of diagnosis.

In our opinion, there are 2 reasons why otologists should be familiar with EPIFs on temporal bone imaging. First, correct identification may prevent unnecessary interventions and follow-up. In this way, the 15 cases (in 10 patients) we described were only incidental findings. Second, in cholesteatoma or cochlear implant surgery, EPIFs may impact the intervention. In our case series, CT imaging suggested a reduced access to the facial recess in 33.3 % (Figure 3). Pertzborn et al. showed that in these cases, safe resection is feasible by careful dissection parallel to the facial nerve, with constant facial nerve monitoring^[1].

CONCLUSION

Epineurial pseudocysts of the intratemporal facial nerve are silent lesions with unknown history. In mastoid surgery, preoperative identification is important to secure a safe and successful procedure and to prevent unnecessary follow-up.

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