

# Ossifying fibroma of the maxilla and sinonasal tract: Case series

Jack J. Liu, M.D.,<sup>1</sup> Lester D. R. Thompson, M.D.,<sup>2</sup> Agnieszka M. Janisiewicz, M.D.,<sup>1</sup> Terry Y. Shibuya, M.D.,<sup>1</sup> David B. Keschner, M.D.,<sup>1</sup> Rohit Garg, M.D.,<sup>1</sup> and Jivianne T. Lee, M.D.<sup>1,3</sup>

## ABSTRACT

**Background:** Head and neck ossifying fibroma (OF) is a rare, benign, locally aggressive, fibro-osseous tumor. The mandible is the most common site of involvement, followed by the maxilla, and, less frequently, the sinonasal cavities, orbit, skull base, and calvarium. In this study, we aimed to expand our understanding of this entity by presenting a case series of OF that involved the maxilla and sinonasal tract.

**Methods:** A multicenter retrospective review was performed on all the patients with a diagnosis of OF from 2004 to 2013. Data were collected with respect to age, sex, clinical presentation, treatment, and outcome.

**Results:** A total of 13 patients were identified. The mean age was 37 years, with a female predominance (69%). The maxillary sinus was most frequently involved site (46%). Eighty-five percent underwent open surgical resection. After a mean follow-up time of 47.3 months, three patients (23%) developed recurrent disease; all of whom were treated with an open approach.

**Conclusion:** OF of the maxilla and sinonasal tract is an uncommon clinicopathologic entity. Although a timely diagnosis may obviate the need for external approaches, open surgical resection is often still necessary for management of extensive lesions. Close follow-up and additional surgery may also be required to treat recurrent disease.

(Allergy Rhinol 8:e32–e36, 2017; doi: 10.2500/ar.2017.8.0190)

Ossifying fibroma (OF) is a benign, fibro-osseous tumor that can occur in the extremities, the head, and the neck.<sup>1</sup> It is histopathologically similar to other fibro-osseous lesions such as fibrous dysplasia and osteomas. According to the World Health Organization, OF is histologically characterized by replacement of normal bone by fibrous tissue that contains varying amounts of mineralized material and cementum (Fig. 1).<sup>2</sup> The etiology of OF is unclear but is thought to be related to multipotential mesenchymal cells of the periodontal ligament or to trauma.<sup>3</sup> OF is a slow-growing, locally aggressive neoplasm. It is typically asymptomatic and does not present until the lesion has become large enough to

cause facial deformity. OF most commonly occurs in the mandible, followed by the maxilla. Although rare, lesions may also occur in the sinonasal tract, orbit, skull base, and temporal bone.<sup>1</sup> Su *et al.*<sup>4</sup> reported that 70% of cases occurred in the mandible and 22% in the maxilla. OF most commonly presents in the second to fourth decade of life and has been reported to be more common in women.<sup>1,5</sup> Sinonasal tract OF reportedly occurs slightly later in life (third and fourth decades) and more commonly in African American women.<sup>6</sup>

A computed tomography (CT) is the preferred imaging modality to characterize these lesions. OF is characterized as a well-defined, radiolucent, unilocular, expansile mass, without bony erosion and scattered radiopaque calcifications (Fig. 2). The definitive management for OF is surgical resection. For mandibular lesions, resection may be accomplished with curettage, whereas extramandibular lesions require endoscopic or open tumor resection.<sup>7</sup> Although OF is common in the mandible and has been frequently described, maxilla and sinonasal tumors are less common. A recent systematic review by Manes *et al.*<sup>8</sup> identified only 137 cases in the literature, a majority of which were derived from isolated case reports. In this review, OF was divided into three histopathologic variants: OF, cemento-OF (COF), and aggressive psammomatoid OF (APOF). To expand our understanding of this rare entity in the maxilla and sinonasal tract, we presented our data and experience from a multi-institutional case series.

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From the <sup>1</sup>Department of Otolaryngology-Head & Neck Surgery, Southern California Permanente Medical Group (SCPMG), Irvine, California, <sup>2</sup>Department of Pathology, Woodland Hills Medical Center, SCPMG, Woodland Hills, California, and <sup>3</sup>Department of Head & Neck Surgery, University of California, Los Angeles (UCLA) Medical Center, Los Angeles, California

No external funding sources reported

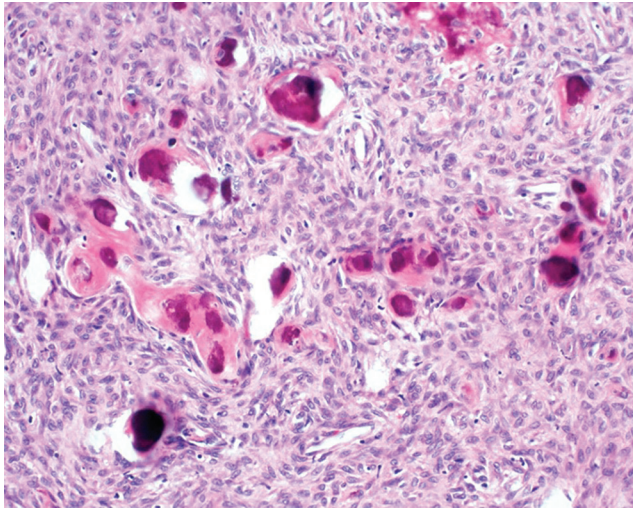
The authors have no conflicts of interest to declare pertaining to this article

Address correspondence to Jivianne T. Lee, M.D., Department of Head & Neck Surgery, University of California, Los Angeles (UCLA) Medical Center, 10833 Le Conte Avenue, 62–132 Center for Health Sciences, Los Angeles, CA 90095

E-mail address: jtlee@mednet.ucla.edu

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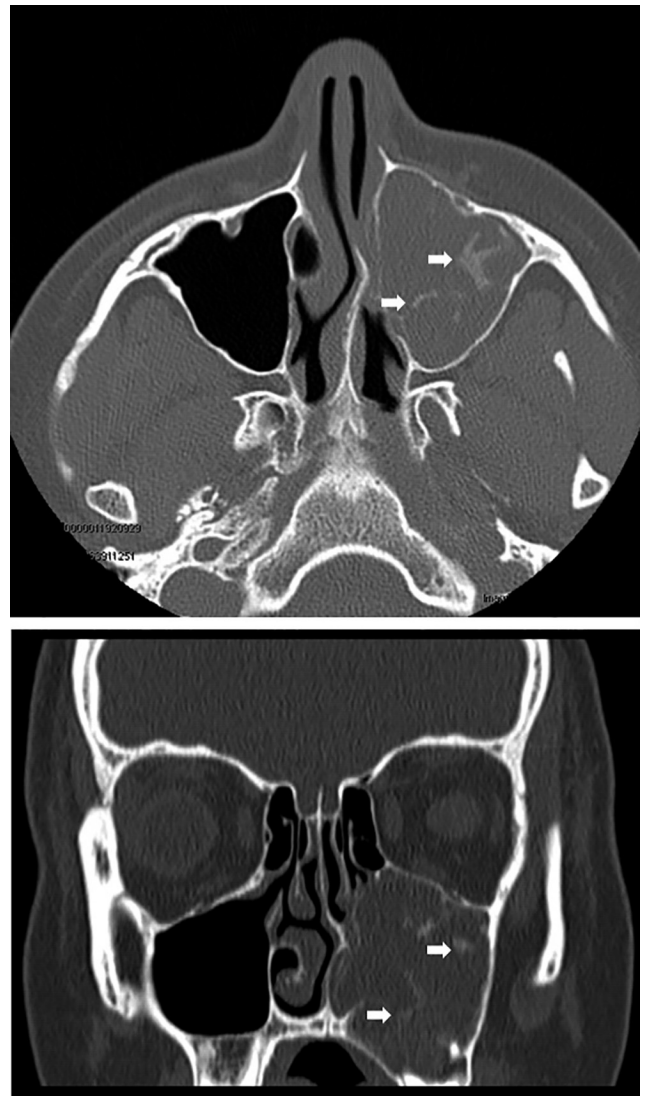
**Figure 1.** This 200× magnification hematoxylin and eosin stained image shows a cellular stroma associated with ossicles of calcium and bone. There is no pleomorphism nor increased mitoses.

## METHODS

This was a multicenter retrospective review from 2004 to 2013 and encompassed seven hospitals (Irvine, Baldwin Park, Downey, Fontana, Sunset, San Diego, Woodland Hills) within the Southern California Permanente Medical Group. This project was approved by the Southern California Permanente Medical Group Institutional Review Board. All patients with a diagnosis of OF in the clinical or pathology data base were included. Only patients with OF in the maxilla, sinonasal tract, orbit, or skull base were included in our review. Patients with incomplete electronic medical records were excluded from this study. All pathology slides were reviewed by a regional head and neck pathologist (L.D.T.). In this study, OFs were classified based on the categorization proposed by Manes *et al.*<sup>8</sup>: OF, COF, and APOF. Electronic medical records were reviewed for age, sex, clinical presentation, treatment approach, and outcomes.

## RESULTS

A total of 23 patients were identified from 2004 to 2013. Nine patients with mandibular OF and one patient with incomplete medical records were excluded from the study. A total of 13 patients from seven hospitals were included in this cohort. The age, sex, location, surgical approach, outcome, and pathologic diagnosis of all the patients included in this cohort are presented in Table 1. The mean age was 37 years, with a range from 6 to 75 years (Table 2). There was a female predominance (69%). The most common location was in the sinonasal tract (69%) and most commonly in the maxillary sinus



**Figure 2.** Axial and coronal computed tomography scan images illustrate an ossifying fibroma of the left maxillary sinus. A well-defined, radiolucent, unilocular, expansile mass is seen without bony erosion. Scattered radiopaque calcifications (arrows) are also present.

(46%). The most common overall presenting symptom was an asymptomatic mass (54%); however, 56% of the patients with sinonasal lesions presented with symptoms such as nasal obstruction or epistaxis. A majority of lesions were removed by using an open approach (85%). The mean follow-up time was 47.3 months. Of the 13 patients, 4 were categorized as having APOF or juvenile active OF. The age of these four patients ranged from 6 to 14 years. The remaining nine patients were categorized as having OF. No COF lesions were noted in this cohort. Three patients had recurrent disease (23%). All the patients initially underwent open resections. Two of these patients had APOF. Each case was discussed in more detail to illustrate the nature of each recurrence.

**Table 1 Demographic information, lesion location, surgical approach, outcomes, and pathology of all the study patients**

Age, Sex y	Location	Year of Surgery	Type of Surgery	Recurrent Disease	Revision Surgery	Pathology
6 F	Left orbital wall	2004	Composite resection, iliac bone graft	Yes	Resection with Medpor reconstruction (2009)	APOF
10 F	Left maxilla	2005	Maxilla curettage	No		APOF
11 M	Right ethmoid sinus	2006	Endoscopic excision	No		APOF
14 F	Left maxillary sinus	2011	Subtotal maxillectomy	Yes	Subtotal maxillectomy (Aug 2012)	APOF
32 M	Right maxilla	2005	Maxillectomy	No		OF
32 M	Left frontal sinus	2010	Coronal approach	No		OF
43 F	Left maxillary sinus	2006	Maxillectomy	Yes	Maxillectomy with STSG (2010)	OF
43 F	Right maxillary sinus	2013	Maxillectomy	No		OF
45 F	Right maxilla	2008	Partial resection of right maxilla bone	No		OF
46 F	Left maxilla	2011	Maxilla bone resection	No		OF
57 F	Left maxillary sinus	2004	Caldwell-Luc	No		OF
67 F	Right frontoethmoid	2005	Endoscopic resection	No		OF
75 M	Left zygoma	2005	Zygomatic bone resection	No		OF

*APOF = aggressive psammomatoid ossifying fibroma; OF = ossifying fibroma; STSG = split-thickness skin graft.*

**Table 2 Summary of cohort data**

Female patients, %	6
Age, mean (range), y	37 (6–75)
Location, no. patients	
Maxilla or zygoma	3
Sinus	9 (6 maxillary sinus)
Orbital wall	1
Resection approach, no. patients	
Open	10
Endoscopic	3
Mean follow-up time, mo	47.3
Recurrences, no. patients	3 (1 endoscopic resection, 2 with APOF)

*APOF = Aggressive psammomatoid ossifying fibroma.*

### Patient 1

A 6-year-old girl presented with an enlarging left lateral orbital mass. A coronal approach was used to excise the mass, including the entire lateral orbital wall, proximal zygomatic arch, and superior aspect of the malar eminence. A full-thickness cranial bone graft and Medpor (Stryker Inc., Kalamazoo, MI) implant were used for reconstruction. A recurrent, enlarging mass was noted at age 10 years. During surgery, the recur-

rent mass was noted to have completely replaced the previous bone graft. The tumor extended onto the orbital floor, skull base, and anterior-most aspect of the zygomatic arch. A bicoronal approach was used, and complete resection was achieved. The patient was noted to have residual disease and was being followed up with serial imaging.

### Patient 4

A 14-year-old girl presented with a left maxillary sinus mass. She underwent endoscopic biopsy and debulking of the tumor. The pathology demonstrated APOF. The patient was taken back to surgery 9 months later for a Caldwell-Luc approach and gross removal of tumor. The patient was noted to have a recurrent mass and was taken back to the operating room 8 months later. A Caldwell-Luc approach was again used. Residual tumor was noted along the inferior, lateral, and anterior maxillary sinus wall. Complete resection was achieved, and there was no evidence of recurrence after 18 months of follow-up.

### Patient 9

A 43-year-old woman presented with a left maxillary sinus tumor. Previous biopsy revealed OF. The patient was taken to the operating room, and a large tumor was found to be occupying the entire maxillary sinus, without bony erosion. An infrastructural maxillectomy was per-



Table 3 Summary of known case series on extramandibular OF

Study	Year of Study	No. Cases	Mean Age, y	OF Type	Surgical Approach (no. patients)	Outcomes	Recurrence Rate, %
Wang <i>et al.</i> <sup>9</sup>	2014	31	25 (median)	OF	Endoscopic (26), open (5)	4 Recurrences in 4–80 mo	13
Wenig <i>et al.</i> <sup>10</sup>	1995	7	33	APOF	Endoscopic (1), open (6)	1 Recurrence in 6 mo to 7 y	14.3
Ledderose <i>et al.</i> <sup>14</sup>	2011	5	49	APOF, COF	Endoscopic (3), declined surgery (2)	No progression in 2 y	None
Lawton <i>et al.</i> <sup>16</sup>	1997	4	28	APOF	Open (3), staged endoscopic (1)	No recurrence in 2 y	None
Harstein <i>et al.</i> <sup>17</sup>	1998	3	16	Psammomatoid OF	Open	No recurrence in 2–10 y	None
Slootweg <i>et al.</i> <sup>18</sup>	1993	3	25	Psammomatoid OF	Limited excision	No recurrence in 2–6 y	None

OF = Ossifying fibroma; APOF = aggressive psammomatoid ossifying fibroma; COF = cemento-ossifying fibroma.

formed, which spared the hard palate. All mucosa within the maxillary sinus was removed, and tooth nos. 13 and 14 were extracted due to suspicion as the tumor nidus. The patient was noted to have a recurrent mass 4 years later. She was taken back to the operating room, and the left hard palate was removed, which completed the maxillectomy. There was no evidence of recurrence after 60 months of follow-up.

## DISCUSSION

OF is a rare entity, particularly in the maxilla and the sinonasal tract. To our knowledge, our cohort of 13 patients was the second largest case series reported and the only multicenter study of extramandibular OF. The largest known case series in the English literature was published by Wang *et al.*<sup>9</sup> in 2014, which consisted of 31 cases of sinonasal OF from Fudan University in Shanghai, China. The recurrence rate was 13% in this case series, and a majority of lesions were resected endoscopically. OF was not classified into its pathologic subtypes in this case series. The next largest series consisted of seven cases of APOF, with a recurrence rate of 14.3%.<sup>10</sup> The remaining case series with at least three patients and published in the English literature are outlined in Table 3. A search of the non-English literature revealed four additional case series, all of which were published in Chinese. Manes *et al.*<sup>8</sup> performed an extensive systematic review of OF in the sinonasal tract. However, a majority of the cases included in the review were derived from individual case reports. There were only four studies that were composed of three or more cases. In addition, as described above, there has actually only been one series with a larger cohort than the one presented.<sup>9</sup> It was a

single center study that was based on a patient population in China, which, unlike the current series, did not classify lesions into its pathologic subtypes. Furthermore, none of the articles in the review were multicenter series.

In our series, OF seemed to be more common in female patients and had a mean age of presentation of 37 years, which was consistent with previous case reports and series.<sup>1,5</sup> Also consistent with previous studies, most patients presented with an asymptomatic mass or facial swelling. In our cohort, patients with paranasal sinus OF tended to present with symptoms such as nasal obstruction or epistaxis. In contrast to Manes *et al.*,<sup>8</sup> APOF seemed to occur exclusively in pediatric patients, consistent with previous reports, in which age <15 years is a hallmark of APOF.<sup>11,12</sup> It should be noted that the cases in this series were performed across multiple centers and by different surgeons over a 10-year period. Therefore, the decision to pursue an endoscopic versus an open approach was left up to the discretion and the experience of the operating surgeons.

Complete removal, particularly in the case of APOF, is the most common recommendation in the literature.<sup>1,13</sup> Ledderose *et al.*<sup>14</sup> indicated that asymptomatic paranasal sinus COF may be managed with a “wait-and-scan” strategy. In their case series, two patients with COF had no progression after 2 years without surgical intervention. Radiation therapy is historically contraindicated due to an increased risk of malignant transformation from 0.4 to 40%.<sup>13</sup>

The recurrence rate in this series was 28% (3/13). Two of the three recurrences were derived from patients with APOF. The literature indicates that APOF

recurrence rates can be as high as 30–58%.<sup>15</sup> Recurrences tended to happen with more extensive tumors, those with APOF, and in pediatric patients. Interestingly, all three recurrences underwent an open approach (2 Caldwell-Luc, 1 bicoronal) as the initial surgery. It is unclear if a different surgical procedure, whether open or endoscopic, would have yielded superior results. Perhaps a more-aggressive initial resection, such as total maxillectomy, could have prevented recurrence, but this must be balanced with causing unnecessary morbidity. One additional recurrence or residual disease was noted after revision surgery in a patient with APOF.

This study was limited by its small sample size and retrospective nature. However, this was the second largest case series in the English literature and the only known multicenter study on OF. It is also important to note that Wang *et al.*<sup>9</sup> consisted of patients in China, which may differ from the patient population in the United States. Furthermore, the series by Wang *et al.*<sup>9</sup> was primarily composed of patients who underwent endoscopic resection as opposed to open removal. The consensus in the literature for management of OF is complete surgical resection; however, subtotal resection should be considered when significant morbidity may occur.<sup>8</sup> There was no evidence that open or endoscopic resection is preferred at this time. Close postoperative surveillance with serial endoscopic examination and radiologic studies is recommended, but the exact duration and means of follow-up required remains unclear.

## CONCLUSION

Data regarding the management and prognosis of sinonasal OF remain elusive. This series illustrated that APOF can recur, particularly in pediatric patients, but larger studies are necessary to corroborate this finding. Although the superiority of endoscopic versus open approaches is still under debate, complete surgical resection when possible with close postoperative follow-up is still recommended.

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