



Retrospective study for diagnosis and management of fibro-osseous lesions affecting paranasal sinuses

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Objectives: *To outline the differentiation, diagnosis and management of different types of fibro-osseous lesions of the maxillofacial region depending on retrospective basis in the previous four years.*

Methods: *A Retrospective study of 10 patients where all patients were diagnosed as having fibro-osseous lesions of the maxillofacial region including fibrous dysplasia, cemento-ossifying fibroma, cherubism, giant cell tumor and Paget disease. There was no age-related contraindication. Patients' age ranged from 10 to 52 years. They were 7 females and 3 males.*

Results: *The mean age was 25.4 years (range: 10 – 52 years). There were 7 females and 3 males. The most frequent location was the maxilla (7 cases). The presenting features were nasal obstruction (7 of 10), facial deformity (6 of 10), proptosis (1 of 10) and decreased vision in 1 case. There were 5 cases of fibrous dysplasia, 2 cases of cement-ossifying fibroma, 1 case of giant cell tumor, 1 case of cherubism and 1 case of Paget disease. Different surgical techniques were used in our study including: endoscopic excision used in 3 cases, endoscopic debulking used in 3 cases, Caldwell-Luc's operation used in one case, and partial maxillectomy used in one case.*

Conclusion: *Radiology is central to the diagnosis of fibro-osseous lesions because the pathology for all of them is similar. Their treatment is highly individualized depending on the type of the lesion and its biological behavior, aggressiveness of the lesion and clinical presentation of the patient. Endoscopy plays an important role in the surgical management of fibro-osseous lesions.*

Keywords: *Fibro-osseous lesions, radiology, endoscopic excision.*

INTRODUCTION

A diverse group of osseous disorders including hereditary or developmental lesions, reactive or dysplastic diseases and neoplasms have been described as benign fibro-osseous lesions.⁽¹⁾ They frequently develop in the craniofacial skeleton and especially in the jaws, the nasal cavity, the paranasal sinuses and the orbit and are mainly characterized by replacement of bone by a connective tissue matrix.⁽²⁾ Fibro-osseous lesions of the maxillofacial bones share overlapping clinical, radiographic and

histopathologic features that may lead to diagnostic confusion and difficulty in differentiation.⁽³⁾

Although the term fibro-osseous lesion had not been included in the WHO's classification of 1992,⁽⁴⁾ these lesions were formally re-classified in 1993⁽⁵⁾ and have been included ever since, based on their biological behavior and histopathology and in agreement with Waldron's recommendations of 1985. Thus fibro-osseous lesions nowadays constitute a group of neoplasms and other tumors related to bone.⁽⁶⁾

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The main clinical symptom is soft tissue swelling and enlargement of the affected bones, which may lead to cosmetic and functional disturbances. The presence of pain, paraesthesia, trismus or dental occlusal findings has been reported, depending on location. Alternatively fibro-osseous lesions may be completely asymptomatic, identified only on routine radiographs⁽⁷⁾ while fibrous dysplasia can be associated with generalized endocrinopathy.⁽⁸⁾

The radiological appearance of fibro-osseous lesions varies depending on the stage of development. In the early stages the lesion is radiolucent and well-defined, while at later stages it changes into radio-opacity with ill-defined borders. The radiographic appearance may thus be either a radiolucent, a mixed radiolucent-radiopaque, a predominantly radiopaque, or ground glass appearance,^(9,10) In craniofacial lesions the bone appearance has been sub classified into three different patterns: pagetoid, cystic or sclerotic.⁽¹¹⁾

The most up to date classification was published by the WHO in 2005.⁽¹²⁾ A much more comprehensive classification has been suggested by Eversole et al in 2008⁽¹³⁾ and this suggests that the classification of these diseases is likely to evolve still further.

WHO Classification 2005:

- Ossifying fibroma
- Fibrous dysplasia
- Osseous dysplasia
- Central giant cell lesion (granuloma)
- Cherubism
- Aneurysmal Bone Cyst
- Simple Bone Cyst

Eversole 2008 Classification (Fig. 1).

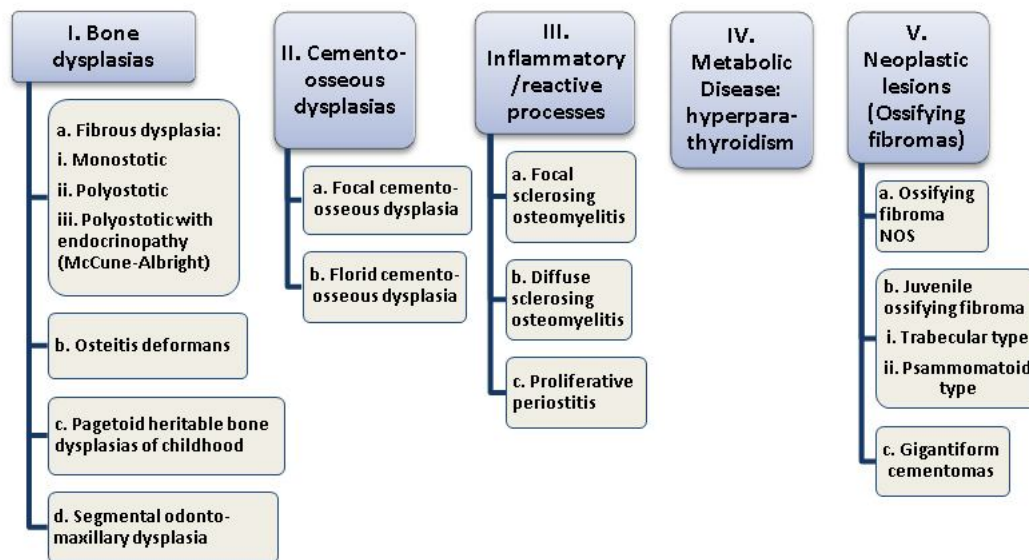


Fig 1. Classification of benign fibro-osseous lesions of the craniofacial complex.

The treatment of fibro-osseous lesions is generally based on their biological behavior and regional aggressiveness. A conservative approach is indicated in some cases, and a more radical approach including a surgical resection may be warranted in others.^(11,14)

PATIENTS AND METHODS

This retrospective study was conducted on 10 patients admitted in the Otorhinolaryngology department, Faculty of Medicine, Cairo University, in the period between 2009 and 2012. All the patients were diagnosed as having fibro-osseous lesions of the maxillofacial region including fibrous dysplasia, cemento-ossifying fibroma, cherubism, giant cell tumor and Paget disease. There was no age-

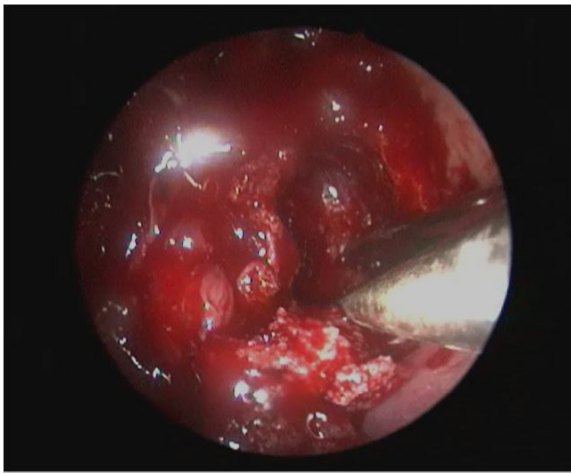
related contraindication. Patients' age ranged from 10 to 52 years. They were 7 females and 3 males.

All the patients were subjected to detailed history, past history and family history of similar conditions. Most of the complaints were deformity, nasal obstruction, headache and eye proptosis. Examination by inspection, anterior rhinoscopy and nasal endoscopy was done. All the cases were subjected to computed tomography (CT), coronal and axial cuts, sometimes with sagittal reconstruction, bone and soft tissue windows with intravenous dye.

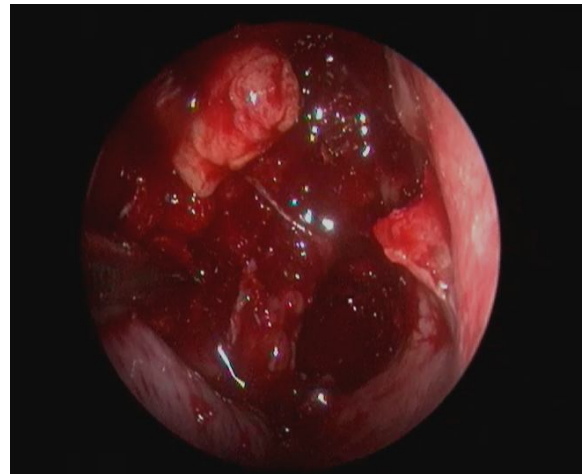
Surgical techniques used in this study included the following: Endoscopic excision, endoscopic debulking, Caldwell- Luc's operation or partial maxillectomy.

All the techniques used were performed under general endotracheal hypotensive anaesthesia with patient in supine position and slight elevation of the patient's head with an angle about 30 degree.

In endoscopic excision (Fig. 2) or debulking (Fig. 3), the surgical steps were performed using a 4 mm straight and angled endoscopes (30° and 45°) in addition to a set of endoscopic instruments and a microdebrider.



A



B

Fig 2a, b. Showing endoscopic excision of ethmoidal and sphenoidal cemento-ossifying fibroma (case no. 2).



A



B

Fig 3a, b. Showing endoscopic debulking of fibrous dysplasia (case no. 8).

Caldwell- Luc's operation was used in extensive maxillary lesions as case number 1 and partial maxillectomy was used in one case (number 6) (Fig. 4).



Fig 4. Right partial maxillectomy (case no. 6).

RESULTS

This retrospective study included 10 patients with fibro-osseous lesions. The mean age was 25.4 years (range: 10 – 52 years). There were 7 females and 3 males. The most frequent location was the maxilla (7 cases). The presenting features were nasal obstruction (7 of 10), facial deformity (6 of 10), proptosis (1 of 10) and decreased vision in 1 case. There were 5 cases of fibrous dysplasia, 2 cases of cemento-ossifying fibroma, 1 case of giant cell tumour, 1 case of cherubism and 1 case of Paget disease.

Table (1) shows the results of this study:

Table 1. Summary for the ten cases included in the current study.

No.	Age	Sex	Location	Radiological characteristics	Clinical features	Histology	Treatment
1	32	M	Lt. maxilla	Ground glass appearance	Facial deformity and left nasal obstruction	FD	Excision via Caldwell-Luc's operation
2	10	F	Rt. sphenoid, ethmoid & maxilla	Circumscribed radio-opaque lesion	Nasal obstruction & headache	COF	Endoscopic excision
3	22	M	Lt. maxilla	Ground glass appearance	Facial deformity and left nasal obstruction	FD	Endoscopic excision
4	52	F	Rt. Ethmoid & frontal sinus	Ground glass appearance	Facial deformity	FD	Endoscopic debulking
5	38	F	Both maxillae & mandible	Multilocular radiolucent areas	Facial deformity	Cherubism	Observation & follow up
6	10	F	Mandible, both maxillae and alveolar ridges	Bilateral asymmetrical opacity	Facial deformity, bilateral nasal obstruction & distortion of teeth	Giant cell tumour	Right partial maxillectomy
7	24	F	Sphenoid & ethmoid	Circumscribed radio-opaque lesion	Nasal obstruction	COF	Endoscopic excision
8	12	F	Lt. ethmoidal, frontal and sphenoidal sinuses	Ground glass appearance	Lt. nasal obstruction & headache	FD	Endoscopic debulking
9	34	M	Maxillo-facial region	Diffuse sclerosis	Asymptomatic	Paget disease	Observation & follow up
10	22	F	Sphenoid, ethmoid, maxillary & frontal sinuses	Ground glass appearance	Facial deformity, proptosis & diminution of vision of lt. eye	FD	Endoscopic debulking & optic nerve decompression

M: Male,

F: Female.

COF: Cemento-ossifying fibroma.

FD: Fibrous dysplasia.

Clinical and radiological data are shown in figures 5-10 as follows:

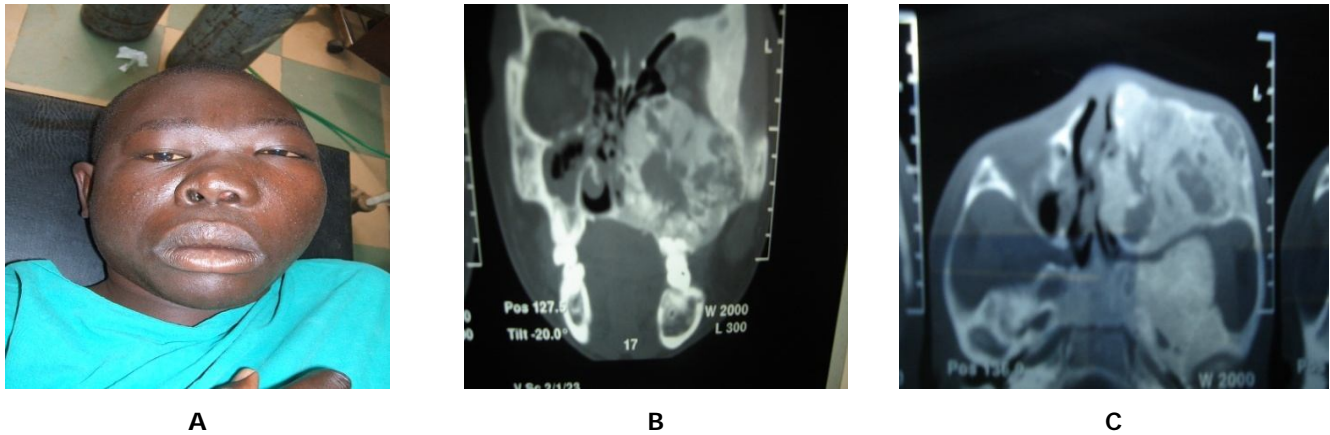


Fig 5a, b, c. Case number 1; fibrous dysplasia with facial deformity and ground glass appearance in the CT scan.

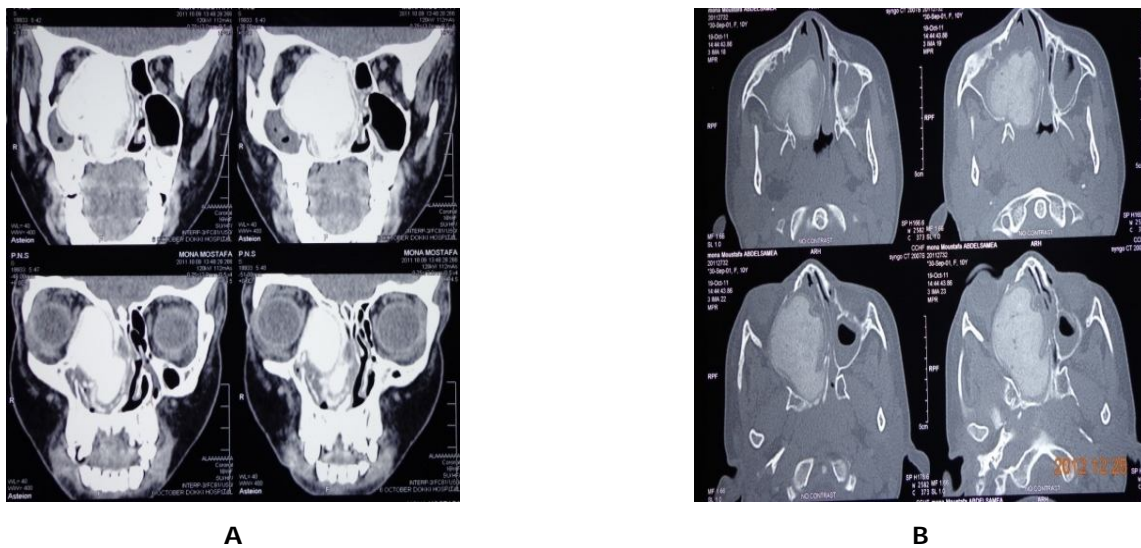


Fig 6a, b. Case number 2; ossifying fibroma in the CT scan.

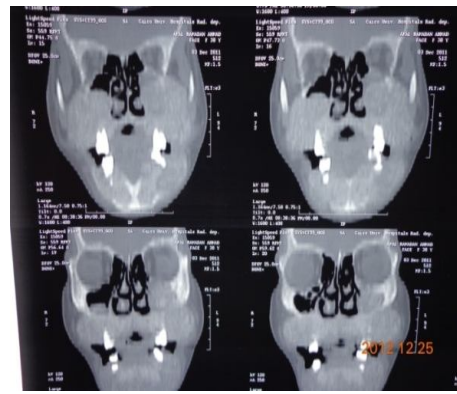


Fig 7. Case number 4; fibrous dysplasia bulging intra cranially and pressurizing the right orbit.

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A



B

Fig 8a, b. Case number 5; Cherubism with bilateral multilocular radiolucent areas within both maxillae and the mandible on both sides.



A



B

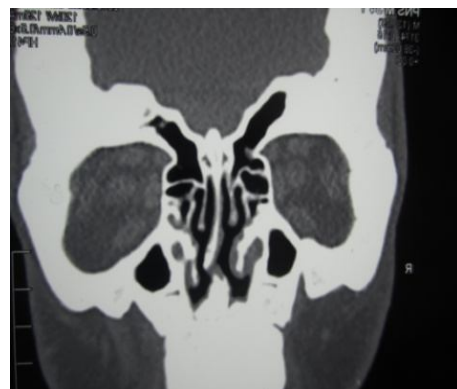


C

Fig 9a, b, c. Case number 6; giant cell tumor with the deformity and the bilateral asymmetrical opacity affecting both maxillae and mandibles.



A



B

Fig 10a, b. Case number 9; Paget disease showing diffuse sclerosis of the maxilla- facial bones.

DISCUSSION

Fibro-osseous lesions of the maxillofacial region constitute a varied group of lesions characterized by replacement of normal bone by tissue composed of collagen and fibroblasts, with variable amounts of a mineralized substance that may be bone, cement or both.⁽¹⁵⁾

These lesions include according to (WHO classification 2005): fibrous dysplasia, ossifying fibroma, osseous dysplasia, giant cell lesion, cherubism, aneurysmal bone cyst and simple bone cyst.

Fibrous dysplasia is a disorder of unknown etiology.⁽¹⁵⁾ In this study, 5 cases of fibrous dysplasia were encountered, 3 females and 2 males. The mean age of presentation of these five cases was 28 years with a range of 12 -52 years. Ogunsalu et al., in 2001 reported a mean age 25.8 years, with a range of 10-47 years⁽¹⁶⁾ while Cardona et al., in 1998 reported a mean age of 49 years with a range of 30-65 years.⁽¹⁷⁾ This lesion is seen more in females than males.⁽¹⁸⁾

Ogunsalu et al., in 2001 stated that the upper maxilla was the most affected location and the lesions tended to manifest as a painless, slow-growing mass.⁽¹⁶⁾ In this study, cases of fibrous dysplasia showed affection of upper maxilla in all the cases.

In our 5 cases of fibrous dysplasia, the lesions presented a radio-opaque and diffuse radiological image. Usually, a mixed image is seen, with areas of increased condensation corresponding to neoformed reticular trabeculae that yield a "ground glass" appearance. The radiological image varies from moderate to diffuse radio-opacity without clear limits.⁽¹⁹⁾

Histologically; the bone is replaced by fibrous tissue, with the gradual appearance of metaplastic bone within the irregular fibrous tissue component. The bone trabeculae appear disperse, in the form of "Chinese characters".⁽¹⁵⁾

The treatment of this lesion tends to be conservative,⁽²⁰⁾ but in our study the 5 cases were subjected to surgical intervention, case 1 → excision through Caldwell-Luc's operation, case 3 → endoscopic excision, while cases^(4,8,10) were subjected to endoscopic debulking.

In the same way ossifying fibroma is a fibro-osseous lesion of unknown etiology but trauma may be an important factor, particularly in the development of cemento-ossifying fibroma.⁽²¹⁾ In our study, the 2 cases of ossifying fibroma (case 2 and 7) gave no history of trauma. From a clinical point of view, division into the cement-ossifying fibroma (COF) and the juvenile ossifying fibroma (JOF) seems to be reasonable.⁽²²⁾

These lesions are more frequent in females than males.⁽²³⁾ In our study the 2 cases were females (cases 2 and 7). And

the age at appearance tends to be variable, though with a certain predominance in the third or fourth decades of life.⁽²⁴⁾ In our study (case 2) had 10 years old and (case 7) had 24 years old.

The mandible is the most common affected location.^(23,25) Ossifying fibromas rarely occur in the paranasal sinuses, e.g. the sphenoid, frontal and ethmoid sinuses, where they are believed to behave more aggressively,⁽²⁶⁾ but in our study the 2 cases affected the paranasal sinuses, (case 2) in the sphenoid, ethmoid and right maxilla, while (case 7) in the sphenoid and ethmoid.

In our study the two cases of cement- ossifying fibroma appear radiologically as mixed, predominantly radio-opaque and well circumscribed images. This coincides with the study done by Dominguez-Cuadrado et al. in 2004.⁽²⁵⁾

The two cases in our study were treated by endoscopic excision but according to MacDonald-Jankowski in 1998, Successful removal can be achieved by curettage, local exeresis and en block resection.⁽²⁷⁾

Because of its locally aggressive behavior and comparatively high vascularisation, the complete surgical resection of an ossifying fibroma of the paranasal sinuses can be challenging, particularly when the frontal skull base or the orbit are involved.⁽²⁶⁾ Consultation of a neurosurgeon or an ophthalmologist might be advisable. Traditionally, microscopical techniques have been used to achieve complete resection.⁽²⁸⁾ In case reports, the endoscopically controlled removal of ossifying fibromas has been described.^(28,29) Based on improved endoscopes and instruments, the further development of operation techniques (e.g. "four-hand-technique")⁽³⁰⁾ and the routine application of precise navigational systems, the limits of the endoscopic approach could be extended over the last decades.⁽³¹⁾ Radiotherapy is considered as ineffective and might even implicate the risk of malignant transformation.⁽³²⁾

In earlier years, concerning fibro-osseous lesions, surgeons were advised to wait until after puberty before operating in the hope that the disease might become arrested at that age. Numerous examples of the disease continuing beyond that period and the continuing dangers to the form and function of young children, so that such concept became unacceptable.⁽³³⁾ The surgical procedures do not adversely alter the growth rates of the normal tissue and do not appear to precipitate the development of malignancy.⁽³⁴⁾ Although conservative surgical resection is ideal in fibro-osseous lesions, a more radical approach is ideal for craniofacial fibro-osseous lesions at unusual locations. This avoids potential skull base complications by compression of disease around foramina and orbital apex. The outcome of surgery in such craniofacial lesions

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can be made favorable with a team approach, i.e., teaming up with the craniofacial surgeons.

Therapeutic optic nerve decompression is less controversial for the treatment of visual loss in fibrous dysplasia. The absolute indications for therapeutic optic nerve decompression are progressive gradual visual loss and occurrence of sudden visual loss within a week's time.⁽³⁵⁾ The patients should also be instructed to report immediately if there are any changes in their vision, including color loss and especially rapid loss of vision. Patients who undergo prophylactic decompression of the optic nerve are difficult to assess for efficacy, because by definition they have no visual loss preoperatively. A lack of development of visual loss postoperatively does not definitely demonstrate efficacy, as it may be possible that they would not have developed visual acuity problems if they were merely observed. Therefore, a large prospective randomized trial study is required to assess the efficacy and safety of prophylactic optic nerve decompression in fibrous dysplasia. Because most of the few reports of decreased vision after decompression occur early in a surgeon's series, prophylactic optic nerve decompression should be done by experienced surgeons for the safety of the procedure.^(35,36) Delayed decompression after 1 week is less likely to improve vision. Combined approaches through intraorbital and intracranial approaches to optical canal are best for decompression.⁽³⁶⁾ In our study, one patient had bilateral proptosis with impaired vision (case 10; fibrous dysplasia). This patient underwent endoscopic debulking with left optic nerve decompression.

CONCLUSION

Fibro-osseous lesions are a heterogeneous group of entities which share similar microscopic features, but exhibit a variety of clinical behavior. Radiology is central to their diagnosis because the pathology for all FOLs is similar. Their treatment is highly individualized depending on the type of the lesion and its biological behavior and clinical presentation of the patient. So, a conservative approach is sufficient in some cases, but in aggressive lesions surgery with complete excision is the treatment of choice in order to decrease recurrence and morbidity. Endoscopy plays an important role in the surgical management of fibro-osseous lesions.

Conflicts of interest

There were no conflicts of interest.

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