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## CASE REPORT

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# Nodular Fasciitis of the Masticatory Space: Magnetic Resonance Imaging Evaluation

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### ABSTRACT

*Nodular fasciitis is a rare benign reactive process of the soft tissues, occurring predominantly in the upper limbs and is less common in the head and neck region. This report describes a histologically proven diagnosis of nodular fasciitis in a 10-year-old boy, who presented with a submandibular swelling. The magnetic resonance imaging findings, its role in this condition, and the differential diagnosis of this uncommon entity are discussed.*

*Key Words:* Child; Fasciitis; Head and neck neoplasms; Magnetic resonance imaging

## 中文摘要

### 咀嚼部的結節性筋膜炎：磁共振成像評估

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結節性筋膜炎是軟組織罕見的良性反應性病變，主要累及上肢，頭部和頸部並不常見。本文報告一名經組織學證實的結節性筋膜炎病例。病例為一名10歲男孩，以下頷下腺區腫脹就診。本文同時討論該病的磁共振成像特徵，及磁共振在診斷及鑑別診斷該罕見病症中的作用。

### INTRODUCTION

Nodular fasciitis (NF) is a benign proliferative lesion characterised by fibroblastic proliferation of soft tissues related to the fascia. The clinical nature of NF has been well understood, with poor understanding of radiological findings.<sup>1</sup> This report describes a 10-year-old boy who presented with a submandibular swelling diagnosed as schwannoma or rhabdomyosarcoma of the masticatory space on magnetic resonance imaging (MRI) following which, the patient underwent surgery and the histology of the specimen was consistent with NF.

The MRI findings of this rare condition are discussed in detail, along with some unusual findings seen in this patient that are usually not seen in NF.

### CASE REPORT

A 10-year-old boy presented in August 2010 with painless swelling in the left submandibular region for 2 months. The swelling increased in size gradually during the 2-month period. No history of trauma or infection was reported. The overlying skin appeared normal (Figure 1), and local examination revealed the mass to

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be soft in consistency; a provisional clinical diagnosis of submandibular gland mass lesion was made and the patient was referred for MRI for further evaluation.

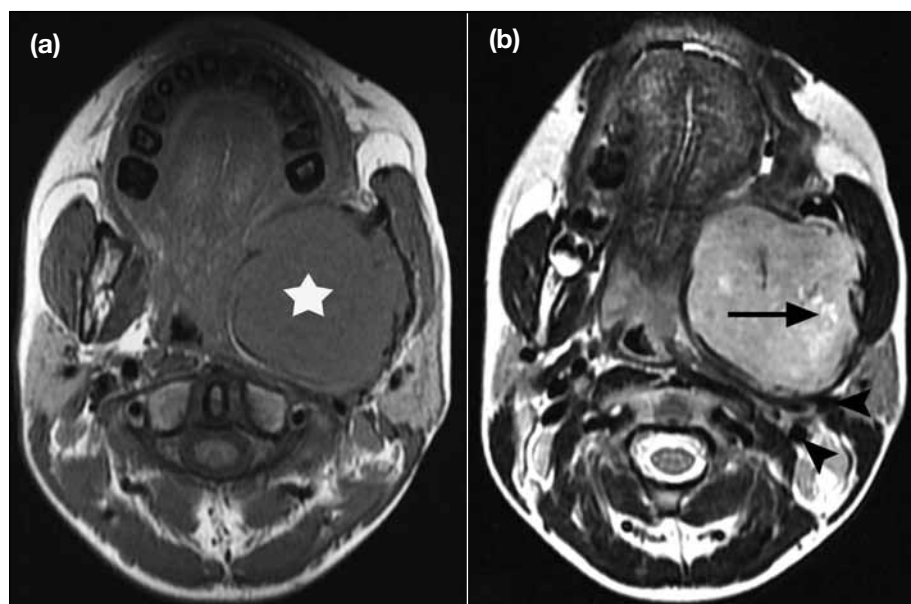
A large well-defined ovoid soft tissue mass was noted in the left masticatory space (Figure 2a), measuring 7 x 5 x 5 cm and causing a pressure effect on the parapharyngeal space with medial displacement of the oropharyngeal wall and posterior displacement



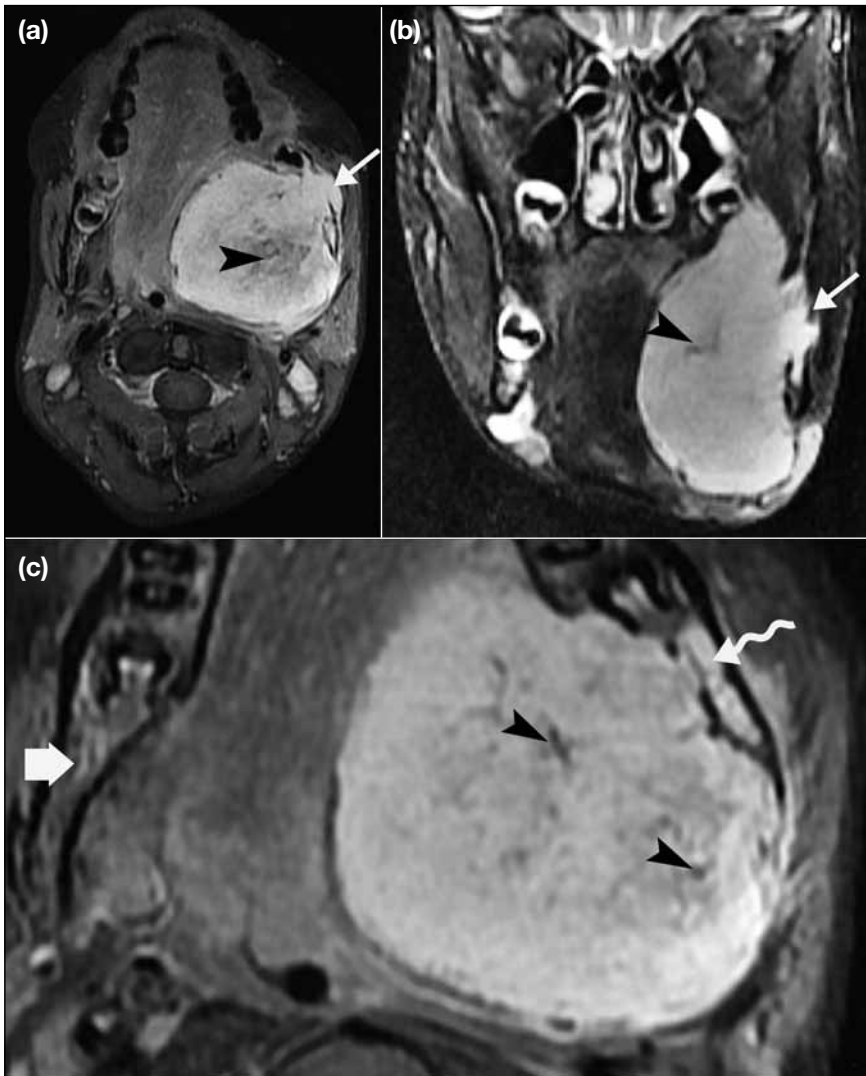
**Figure 1.** Submandibular swelling on the left side of the face.

of the carotid vessels. The mass appeared isointense to adjacent muscle on T1-weighted image and hyperintense to adjacent muscle on T2-weighted image (Figure 2b). The mass showed multiple small hyperintense areas at the centre, representing mucoid-filled cystic spaces. The internal jugular vein at the level of the mass was not visualised as it was compressed by the mass. Post-contrast fat-suppressed T1 image showed marked enhancement of the lesion with multiple small non-enhancing hypointense areas at the centre representing fluid-filled mucoid cystic spaces (Figures 3a and 3b). The mass had caused erosion of the adjacent ramus of the mandible with transcortical extension and had extended to involve the inferior alveolar canal on the left side (Figure 3c). Post-contrast fat-suppressed T1 image (Figure 4a) and short T1 inversion recovery image (Figure 4b) showed a hyperintense mass with a normally appearing body of the mandible showing normal marrow signal intensity. In view of the location, age of the patient, morphology of the lesion, and enhancement pattern, a provisional diagnosis of malignant schwannoma or rhabdomyosarcoma of the masticatory space was made.

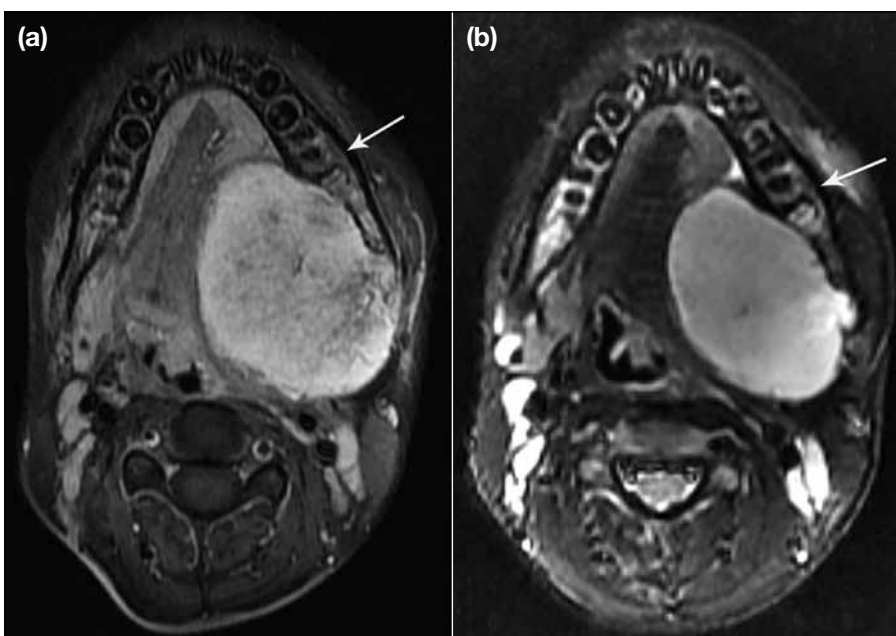
The patient underwent surgery, during which the mass was found to be well defined and was easily removed. There was erosion of the adjacent ramus of the mandible. Healing was satisfactory. Histopathology of the specimen showed myofibroblasts in myxoid stroma, uniform in size and shape with pale-staining nuclear units and prominent nucleoli with few



**Figure 2.** Magnetic resonance (a) axial T1-weighted and (b) T2-weighted images showing a large well-defined ovoid soft tissue mass in the left masticatory space (star) isointense to adjacent muscle on T1 and hyperintense to adjacent muscle on T2 images causing a pressure effect on the parapharyngeal space, with medial displacement of the oropharyngeal wall and posterior displacement of the carotid vessels (arrowheads). The mass shows multiple small hyperintense areas at the centre (arrow) representing mucoid-filled cystic spaces. The internal jugular vein at the level of the mass is not visualised as it is compressed by the mass.



**Figure 3.** Magnetic resonance (a) axial and (b) coronal post-contrast fat-suppressed T1-weighted images showing marked enhancement of the lesion with multiple small non-enhancing hypointense areas at the centre of the mass representing fluid-filled mucoid cystic spaces (arrowheads). The mass has eroded the adjacent ramus of the mandible (arrows) with transcortical extension and (c) extends to involve the inferior alveolar canal on the left side (curved white arrow). The canal on the right side appears normal (short white arrow).



**Figure 4.** Magnetic resonance (a) post-contrast fat-suppressed axial and (b) short T1 inversion recovery images showing a hyperintense mass with a normal-appearing body of the mandible with normal marrow signal intensity (arrows).

lymphocytes and microhaemorrhages. No atypia was seen. Immunostaining technique was not performed. On the basis of the presence of myofibroblasts in the myxoid stroma with lymphocytic infiltration and microhaemorrhages without atypia (Figure 5), a diagnosis of NF was made.

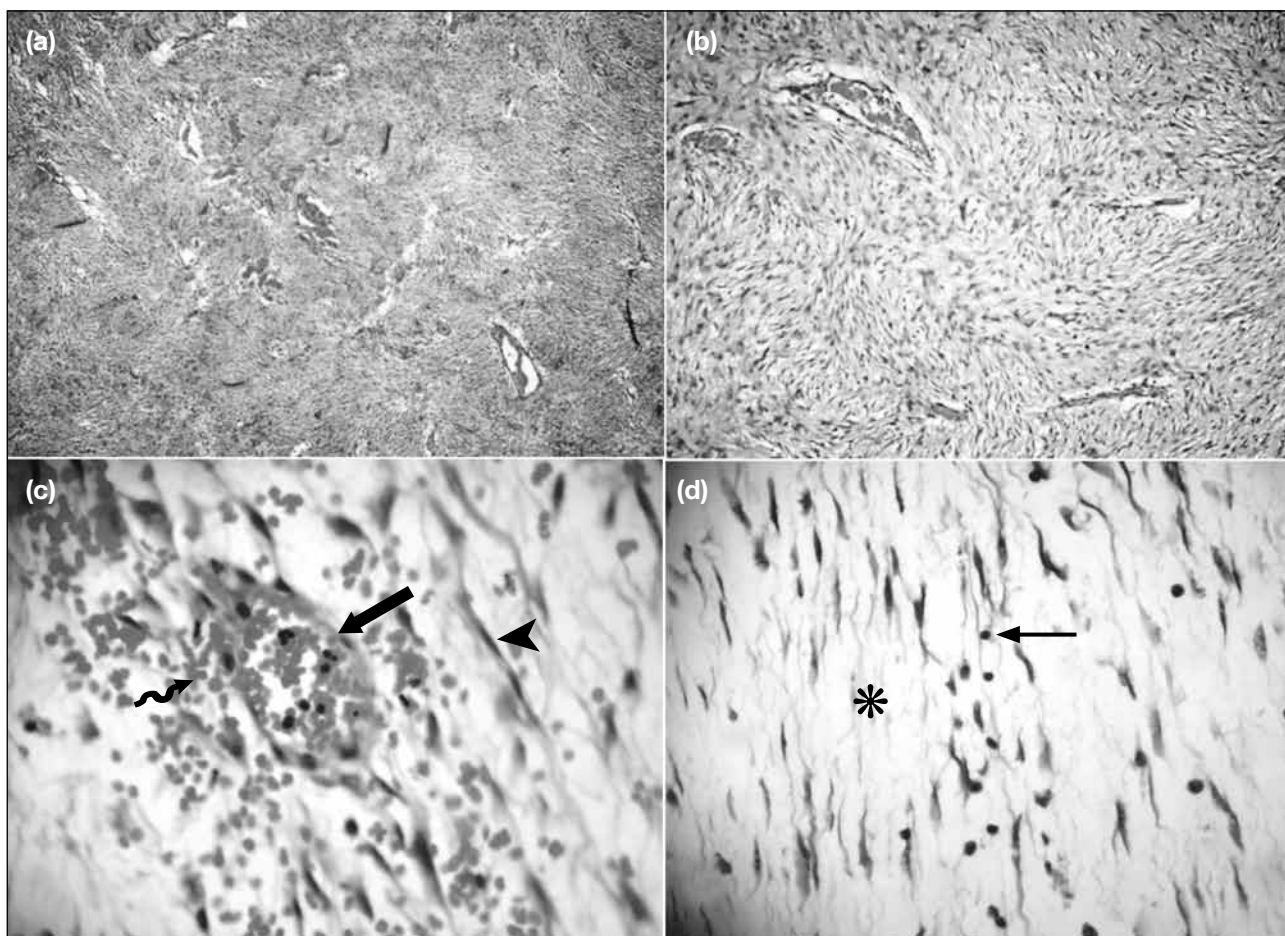
The patient has been reviewed regularly, and there is no sign of recurrence after 10 months.

## DISCUSSION

NF is a rare entity, which most often affects the trunk and upper limbs.<sup>2</sup> NF can be found anywhere in the body, with equal incidence in men and women. The most common sites noted are the upper extremities (48%), trunk (20%), head and neck (15-20%), and lower extremities (15%). NF can affect any age group, but is most commonly seen in patients aged between 20

and 40 years. Less than 20% of cases occur in patients younger than 20 years, and only 5% in patients older than 70 years. Lesions vary between 0.5 and 10 cm in size, and larger lesions are rare.<sup>3</sup> NF is rare in the head and neck region, but occurrence in this location is more common among children.<sup>4</sup>

The exact pathogenesis of this rare entity is not known, although it is thought to be a self-limiting condition of either reactive or inflammatory aetiology and may be triggered by trauma in some instances. Lesions are well defined and soft in consistency, and adhere firmly to underlying structures. Lesions are usually asymptomatic, but pain may be present in some patients.<sup>2</sup> There are three types of NF depending on their location: subcutaneous, intramuscular, and intermuscular (fascial). The subcutaneous type occurs 3 to 10 times more commonly than the other subtypes and presents as a



**Figure 5.** Histopathological staining showing (a) feathery tissue culture-like appearance (H&E; original magnification, x 4); (b) loosely arranged in C and S bundles (H&E; original magnification, x 10); (c) elongated nuclei (arrowhead) with small thin-walled blood vessel (arrow) with extravasated blood (wavy arrow) [H&E; original magnification, x 40]; and (d) loosely arranged myofibroblasts in myxoid stroma (\*) with uniform and scattered lymphocytes (arrow) [H&E; original magnification, x 40].

subcutaneous nodule. The intramuscular type is most likely to mimic a soft tissue malignancy because of its size and location. The intermuscular type is not well circumscribed and grows along the fascial planes. Rare subtypes are intravascular and intradermal forms. A relationship exists between these subtypes and the age of the lesion in that the younger the lesion, the more myxoid component is present. If the lesion is mature, then fibrotic component predominates.<sup>3</sup> On histology, NF is abundantly cellular, consisting of immature fibroblasts with prominent nucleoli and spindle- or stellate-shaped cells in myxoid stroma associated with lymphocytic infiltration and microhaemorrhages.<sup>5</sup>

This specimen showed myofibroblasts, which were immature, spindle-shaped, and uniform in size and shape, with pale-staining nuclear units and prominent nucleoli. Lymphocytic infiltration was sparse with microhaemorrhages. The tumour was well circumscribed, but showed bony erosion and no atypia was seen. Since the lesion was of a short duration, the inflammatory component was less and myxoid stroma was predominant. Presence of myofibroblasts in myxoid stroma with lymphocytic

infiltration, microhaemorrhages, and absence of atypia were the features consistent with NF. Although immunohistochemical examination was not performed for this patient, an accurate diagnosis of NF should include immunohistochemical data for myogenic markers such as alpha-smooth muscle actin, muscle-specific actin, desmin, myogenic determination factor, myogenin, S-100, and Ki-67.

Some of the differential diagnoses to be considered include schwannoma,<sup>6</sup> sarcomas (fibrosarcoma, rhabdomyosarcoma, and leiomyosarcoma),<sup>6</sup> chondrosarcoma,<sup>6</sup> soft tissue phlegmon,<sup>7</sup> and non-Hodgkin's lymphoma<sup>8</sup> (Table), as follows:

- Schwannoma: Antony A or B pattern or wavy nuclei are noted in this condition, but not seen in NF.
- Intramuscular myxoma: myxoma is paucicellular and poorly vascularised whereas NF is richly cellular with abundant capillaries.
- Malignant fibrous histiocytoma and other sarcomas: no pleomorphism, hyperchromasia, or atypical mitosis, and cells are uniform spindle or stellate myofibroblasts.

**Table.** Differential diagnosis based on magnetic resonance imaging findings.

	T1-weighted	T2-weighted	Post contrast	Differentiating points
Nodular fasciitis	Hypointense	Hyperintense	Strong enhancement with non-enhancing areas	<ul style="list-style-type: none"> <li>• Subcutaneous type: well-defined and surrounded by fat</li> <li>• Fascial (intermuscular) type: thickening of fascial septa is present</li> <li>• Intramuscular type: ill-defined margins and can invade or destroy adjacent structures, including the bone; mimics soft tissue malignancy</li> </ul>
Schwannoma	Iso- to hypo-intense with few hyperintense areas representing haemorrhage	Iso- to hyper-intense	Homogeneous or heterogeneous enhancement with cystic or necrotic areas	<ul style="list-style-type: none"> <li>• Associated with atrophy of masticatory muscle</li> <li>• Smooth widening of foramen ovale</li> </ul>
Fibrosarcoma or rhabdomyosarcoma	Iso- to hypo-intense with 30% showing high signal due to haemorrhage	Heterogeneously hyperintense to muscle with poorly defined margins	Heterogeneous enhancement on post-contrast	Findings mimic intramuscular variant of nodular fasciitis and is difficult to differentiate, but perineural spread is common and intracranial extension is also noted in some cases of rhabdomyosarcoma
Chondrosarcoma	Homogeneous intermediate signal, cartilage or calcification make signal heterointense	Increased signal intensity either homogeneous or heterogeneous depending on type of calcification	Enhancement depends on tumour grade; the higher the grade, the more the enhancement	Calcification of variable pattern with presence of rings and crescents or amorphous types of calcium
Non-Hodgkin's lymphoma (extra nodal)	Isointense to muscle	Hyperintense	Strong enhancement on post-contrast study	Findings of non-Hodgkin's lymphoma may mimic intramuscular variant of nodular fasciitis and it may be difficult to differentiate on magnetic resonance imaging, but extranodal non-Hodgkin's lymphoma in head and neck region is usually located in Waldeyer's ring and 40-60% of patients have systemic non-Hodgkin's lymphoma

Variations in signal intensities are noted on MRI, and these variations depend on variability in cellularity, amount of collagen content, cytoplasm, water content of the extracellular space, and vascularity of the lesion.<sup>9</sup> Lesions are hypointense on T1 images and hyperintense on T2 images<sup>10</sup>; subcutaneous lesions are well defined and surrounded by fat, and the appearance varies according to the histological component of the lesion.<sup>11</sup>

According to the literature, deep-seated lesions are of intramuscular type, with ill-defined margins, and can invade or destroy adjacent structures, including bone.<sup>3</sup> In cases of fascial-based mass lesion, thickening of the fascial septa is an MRI sign to raise the possibility of NF.<sup>9</sup> On post-contrast study, these lesions show enhancement with variations in enhancement pattern.<sup>10</sup> Enhancement is due to the prominent capillary network and compact cellularity of the lesion, with few non-enhancing cystic areas within the lesion that appear hyperintense on T2 images, and are mainly due to fluid-filled mucoid spaces. There is a relationship between the signal intensities on T2 images and histological subtypes of the lesion. Signal intensity of lesions with myxoid or cellular histology is higher than that of muscle, whereas lesions with fibrous histology appear hypointense to the surrounding muscles on all sequences. Presence of abundant collagen and reduced cellularity leads to reduction in signal intensity in the fibrous type.<sup>3</sup> NF involving the head and neck region are partially embedded in the associated adjacent muscle, sometimes with adjacent fascial thickening. These lesions appear benign with moderate-to-strong enhancement on computed tomography and MRI, but sometimes show aggressive behaviour in the form of local bony destruction and intracranial extension.<sup>1</sup>

The lesion in this patient was deep-seated in the masticator space, partially embedded in the adjacent muscle. The lesion was well-defined with smooth walls showing strong contrast enhancement with few non-enhancing cystic areas, representing fluid-filled mucoid spaces. The lesion showed aggressive behaviour in the form of erosion of the adjacent ramus of the mandible.

The radiological differential diagnosis depends on the anatomical locations and radiological findings. The specific diagnosis is difficult, and it is often not possible to diagnose even with MRI. It is difficult to differentiate between benign and malignant soft tissue lesions in some patients.

Subcutaneous lesions resolve spontaneously within a few weeks but, if they do not resolve, surgical excision is performed.<sup>11</sup> Superficial lesions are treated by simple excision and deeper ones by wide excision. Recurrence of tumour is rare, and any recurrence may warrant pathological review to exclude a misdiagnosis of malignant sarcoma as NF.

## CONCLUSION

NF is a rare condition, with about 15% occurring in the head and neck region. There are three types of NF depending on their location: subcutaneous, intermuscular (fascial), and intramuscular. Sometimes, the lesions present without a history of trauma. Although benign, lesions can grow up to 10 cm in size, even in children, and have local aggressive features. There is no pathognomonic clinical or radiological sign suggestive of NF, and the condition needs clinical and radiological correlation to arrive at a diagnosis. NF should always be included in the differential diagnosis of soft tissue masses in the head and neck region, especially in patients presenting with a recent history of trauma followed by a rapidly growing mass.

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