

# World Journal of *Clinical Cases*

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Observational Study

# Imaging characteristics of orbital peripheral nerve sheath tumors: Analysis of 34 cases

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## Abstract

### BACKGROUND

Peripheral nerve sheath tumors (PNSTs), a rare group of neoplasms in the orbit, comprise only 4% of all orbital tumors. At present, there are very few studies detailing the features of these tumors identified using imaging technology.

### AIM

To compare the differences in location, morphology, magnetic resonance imaging (MRI) signal intensity/computed tomography (CT) value, and enhancement degree of tumors of different pathological PNSTs types.

### METHODS

Clinical, pathological, CT, and MRI data were analyzed retrospectively in 34 patients with periorbital sheath tumors diagnosed using histopathology from January 2013 to August 2021.

### RESULTS

Among 34 cases of orbital peripheral nerve sheath tumors, 21 were schwannomas, 12 were neurofibromas, and 1 was a plexiform neurofibroma. Common clinical symptoms presented by patients with these types of tumors include eyelid swelling, exophthalmos, and limited eye movement. Schwannomas mostly occur



in the intramuscular space with small tumor volume and rare bone involvement. Neurofibromas develop in the extrapyramidal space with larger tumor volume and more bone involvement. Radiologically, schwannomas and neurofibromas are characterized by regular morphology and uneven density and signal. One case of plexiform neurofibroma showed tortuous and diffuse growth along the nerve, with a worm-like appearance on imaging.

### CONCLUSION

Different pathological types of orbital peripheral nerve sheath tumors have unique imaging characteristics. Comprehensive consideration of the patient's clinical and imaging manifestations is of great value in the diagnosis of orbital peripheral nerve sheath tumors.

**Key Words:** Periorbital nerve sheath tumor; Schwannoma; Neurofibroma; Plexiform neurofibroma; Imaging features

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**Core Tip:** We analyzed clinical, pathological, computed tomography (CT), and magnetic resonance imaging (MRI) data retrospectively in 34 patients with periorbital sheath tumors diagnosed using histopathology during more than 7 years. The differences in location, morphology, MRI signal intensity/CT value, and enhancement degree of tumors of different pathological types were compared. Radiologically, schwannomas and neurofibromas are characterized by regular morphology and uneven density and signal. One case of plexiform neurofibroma showed tortuous and diffuse growth along the nerve, with a worm-like appearance on imaging.

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### INTRODUCTION

In 1768, Akenside first published a scientific description of a patient with multiple tumors involving the peripheral nerves[1]. Over ten decades later, von Recklinghausen demonstrated that neurofibromas belong to a single nerve[2]. In 1910, neurinoma was histologically independent of neurofibromas and was later named schwannoma after confirming that the tumor originated in Schwann cells[3]. An in-depth study subdivided and reclassified peripheral nerve tumors that develop from the neural crest and neuroectoderm as peripheral nerve sheath tumors (PNSTs). According to the 2020 World Health Organization classification of soft tissue tumors, PNSTs are divided into categories, including neurilemmomas (schwannomas), neurofibromas, plexiform neurofibromas, hybrid nerve sheath tumors, nerve sheath myxomas, meningiomas, and malignant peripheral nerve sheath tumors[4]. Malignant PNSTs account for an extremely low proportion of cases and have a special gross appearance of fusiform, fleshy, white mass with degeneration, and secondary hemorrhage. More than 95% of PNSTs are benign, and schwannomas are the most common benign PNST that appear heterogeneous because of their histological composition[5].

The infraorbital nerve tissue is rich, including the optic nerve entering the middle cranial fossa through the optic nerve canal, oculomotor nerve, trochlear nerve, abductor nerve, the first branch of the trigeminal nerve entering the orbit through the supraorbital fissure (eye meridian), the supraorbital nerve from the supraorbital fissure to the forehead, and the infraorbital nerve entering the orbit through the infraorbital fissure[6]. Except for the optic nerve, all infraorbital nerves are peripheral nerves. Orbital PNSTs have a low incidence rate, accounting for only approximately 4% of all orbital tumors. They are thought to originate from sensory nerves and are frequently located in the superior and medial orbital compartments. However, PNSTs rarely invade the orbit and ocular adnexa[7]. Orbital PNSTs are often characterized by variable tumor locations, nonspecific clinical symptoms, occasional diagnostic dilemmas, and challenging treatment plans[8]. Although it may be difficult to differentiate orbital masses based on imaging data, computed tomography (CT) and magnetic resonance imaging (MRI) features can still provide clues for diagnosis[9]. For example, localized tumors on CT often demonstrate smooth margins, round, ovoid, homogenous density, or lobulation[10].

In this study, we summarized the imaging, pathological, and clinical characteristics of 34 patients in detail, which provides valuable references and ideas for the differential diagnosis of this disease.

## MATERIALS AND METHODS

### Study participants

We recruited 34 histologically-diagnosed cases of PNSTs admitted to the Department of Ophthalmology, Xi'an People's Hospital (Xi'an Fourth Hospital), Shaanxi, China, from January 2013 to August 2021. Specifically, 21 cases were schwannomas, 12 cases were neurofibromas, and 1 case was plexiform neurofibroma. Three groups were defined according to tumor type. None of these patients had undergone any kinds of therapy or had any other malignancy. All patients underwent imaging examinations: 28 patients underwent CT scan, MRI-plain scan, and enhanced scans; two patients received both MRI-plain scan and enhanced scan; 2 patients underwent MRI-plain scan only; and 2 patients received CT scan only.

Our study was approved by the Institutional Committee for Research Involving Human Subjects of the Xi'an People's Hospital (Xi'an Fourth Hospital). Informed consent forms listing relevant information needed to be collected were signed and obtained from the participants. Demographic and personal data, including gender, age, and clinical symptoms, were collected from outpatient medical records. We retrospectively analyzed the clinical and imaging features of these patients.

### Imaging examination

CT scanning using a 16-row spiral CT machine (Siemens, SomAToM Emotion, Germany). The scanning parameters were as follows: plain scan slice thickness, 2.0 mm; slice increment, 2.0 mm; and reconstruction slice thickness, 0.75 mm. CT scans of four patients were performed on a 64-slice CT machine (GE Revolution EVO, United States). The detector width was 20.0 mm, and the slice thickness, pitch, and reconstruction slice thickness were 2.5 mm, 0.969 mm, and 0.75 mm, respectively. Sagittal and coronal reconstructions were performed.

MRI scans of all patients were performed using a 3.0 magnetic resonance scanner (GE, Signa HDxt, United States) and a skull 8-channel phased array coil. Plain orbital MRI scan sequences included T1-weighted imaging (T1WI) axial repetition time (TR)/echo time (TE; TR/TE: 400 ms/10 ms), T2-weighted imaging (T2WI)/fat saturation (FS) axial (TR/TE: 2800 ms/70 ms), T2WI/FS oblique sagittal (TR/TE: 1500 ms/70 ms) and T2WI/Short TI Inversion Recovery (STIR) coronal (TR/TE: 6600 ms/40 ms). The orbital MR enhancement sequence included the T1WI/FS axial position, oblique sagittal position (TR/TE: 500 ms/10 ms) and coronal position (TR/TE: 450 ms/10 ms). The slice thickness was 2 mm, and the slice increment was 1 mm. Meglumine gadopentetate (Grant No. j20171008, Bayer), a contrast agent, was rapidly injected at a dosage of 0.2 mL/kg.

### Statistical analysis

Two senior imaging physicians used uniform criteria to evaluate the images. If the results were inconsistent, a consistent diagnosis was reached through consultation. Physicians evaluated the following characteristics: tumor location (internal and external space of the muscle cone), shape (regular or irregular), size, CT value, MRI signal intensity (taking cerebral gray matter as reference), signal uniformity (uniform or non-uniform), enhancement degree (mild, moderate, obvious, non-enhancement), and enhancement pattern (uniform or non-uniform).

Data were collected using Microsoft Excel, and statistical analyses were performed using IBM SPSS Statistics for Windows version 26.0. The count data are expressed as cases and percentages (%), and a chi-square test was performed to compare the variables among the three groups. We adopted a continuity-adjusted formula for the chi-square test if the value of the expected cases in one cell was greater than or equal to 1 but less than 5. Fisher's exact test was used if a cell had few expected cases (*i.e.*, < 1) in the table. We considered a 2-tailed *P* value < 0.05 as statistically significant. Since there was only one case of plexiform fibroma, we fused the plexiform case to the neurofibroma group to compare the patients' general characteristics.

## RESULTS

### Population characteristics

In this study, we recruited 34 patients with PNSTs, including 21 with schwannoma, 12 with neurofibroma, and 1 with plexiform neurofibroma. Their ages ranged from 18 to 77 years old. **Table 1** presents the demographic and clinical characteristics of the study population. The mean age of the schwannoma patients ( $47.33 \pm 3.602$  years) was not significantly different from that of the patients with neurofibroma ( $41.77 \pm 4.595$  years). There were 17 male and 17 female patients, of whom the male-to-female ratio of schwannoma was approximately 4:3, and that of neurofibroma was 5:8. The most common clinical symptoms of the three tumors were eyelid swelling, exophthalmos, and limited eye movement, and the difference in prevalence based on tumor type was not statistically significant (all *P* > 0.05).

**Table 1 Characteristics of 34 peripheral nerve sheath tumors patients**

	Schwannoma (n = 21) (%)	Neurofibroma + plexiform neurofibroma (n = 13) (%)	P value
Gender			0.290
Male	12 (57.1)	5 (38.5)	
Female	9 (42.9)	8 (61.5)	
Age (mean ± SD, yr)	47.33 ± 3.602	41.77 ± 4.595	0.347
Clinical symptoms			
Exophthalmos	14 (66.7)	9 (69.2)	1.000
Limited eye movement	11 (52.4)	7 (53.8)	0.934
Eyelid swelling	14 (66.7)	7 (53.8)	0.455
Decreased vision	4 (19.0)	1 (7.7)	0.682
Dizzy	6 (28.6)	2 (15.4)	0.642
With NF-I	0 (0.0)	4 (30.8)	0.015 <sup>a</sup>

<sup>a</sup>P < 0.05.

NF-I: Neurofibromatosis type I.

### Imaging characteristics

**Table 2** displays differences in imaging findings between the group of schwannoma patients and the neurofibroma group. Generally, statistically significant differences in tumor location and bone involvement were observed between two groups, while other radiological features, such as tumor morphology, density, and signal uniformity, were similar. Concretely speaking, the imaging manifestations of the two tumors had regular morphology, and uneven density and signal. Schwannomas mostly occur in the intramuscular space and have relatively small volume, thus having uncommon bone involvement, as shown in **Figure 1**. However, neurofibroma, often having large tumor volume and adjacent bone compression, mostly occurs in the extrapyramidal space (**Figure 2**). As for the case of plexiform neurofibroma, MRI examination showed that the tumor grew diffusely and tortuously along the nerve; multiple slightly long T1 and slightly long T2 signal nodules were found in the right orbit. Further, small flake and strip T2 signal shadows were seen in the T2WI lesions, with a diameter of about 4 to 5 mm. Through the infraorbital hole and supraorbital fissure, the tumor grew into the intracranial, deep cervical, and subclavian spaces (**Figure 3**).

## DISCUSSION

Although multiple studies have described the clinical or histological features of orbital PNST, the literature still lacks a systematic imaging description of these tumors[11,12]. We performed a comprehensive radiological review of several PNSTs from the orbit in the context of the current classification framework.

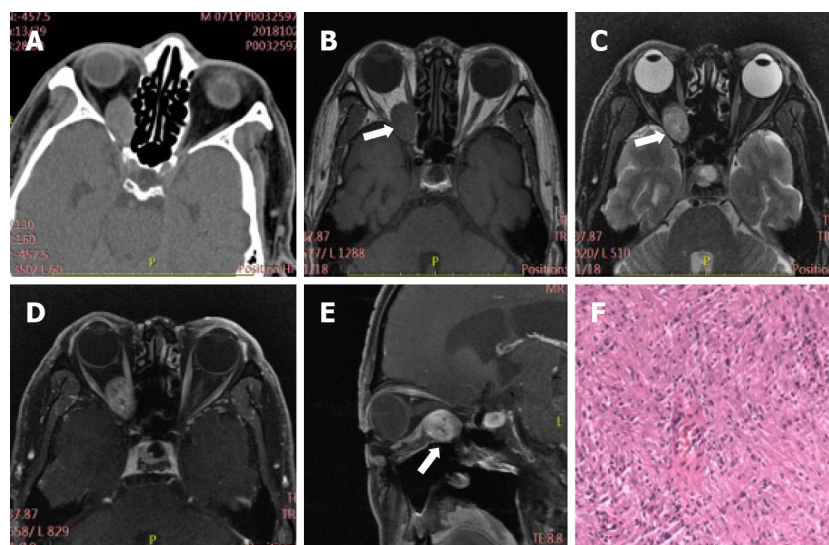
PNSTs often occur in the neck and head but are infrequent in the orbit. Based on five large studies of biopsy-proven orbital tumors, Sweeney *et al*[10] reported the occurrence probability of PNSTs in different orbital tumors: schwannoma 0.7%-2.3%, neurofibroma 0.4%-3.0%, malignant PNST 0%-0.2%. Unlike malignant PNSTs, which have a peak incidence in the seventh decade, benign orbital PNSTs are considered tumors of adulthood, with the exception of plexiform neurofibromas, of which approximately 50% are diagnosed in early childhood[13,14]. In our study, the mean age of patients with schwannoma and neurofibroma was 47.33 ± 3.602 years and 42.17 ± 4.977 years, respectively. The patient with plexiform neurofibromas was 37 years old; only one case was included in the study because of the low prevalence of this type of PNST. Generally, the age of onset in these patients is consistent with the epidemiological characteristics of this disease.

The molecular etiology of PNSTs remains unclear[15]. Previous studies have demonstrated that loss of the tumor suppressor gene *NF-1* (17q 11.2) is related to 28% of neurofibromas and almost all plexiform neurofibromas, but not neurilemmomas[16]. In our study, none of the 21 schwannoma cases were accompanied by *NF-1*, although three neurofibroma cases (25.0%) and only one plexiform neurofibroma case had *NF-1*.

Schwannomas arise from myelin-producing Schwann cells and grow principally *via* cell hyperplasia, often showing a predominance of spindle-shaped Schwann cells[17]. Pathologically, a schwannoma is a smooth/unsmooth tan or yellow mass with occasional hemorrhage, calcification, or atypical cystic

**Table 2 Comparison of imaging features between schwannoma and neurofibroma**

Imaging features	Schwannoma (n = 21) (%)	Neurofibroma (n = 12) (%)	Chi-square value	P value
Location (Intramuscular space)			11.933	0.000 <sup>a</sup>
Yes	19 (90.5)	3 (25.0)		
No	2 (9.5)	9 (75.0)		
Location (extrapyramidal space)			11.933	0.000 <sup>a</sup>
Yes	2 (9.5)	9 (75.0)		
No	19 (90.5)	3 (25.0)		
Regular morphology			0.002	0.968
Yes	16 (76.2)	10 (83.3)		
No	5 (23.8)	2 (16.7)		
Bone involvement			5.085	0.024 <sup>a</sup>
Yes	3 (14.3)	7 (58.3)		
No	18 (85.7)	5 (41.7)		
Homogeneous density/signal intensity			0.000	1.000
Yes	6 (28.6)	3 (25.0)		
No	15 (71.4)	9 (75.0)		

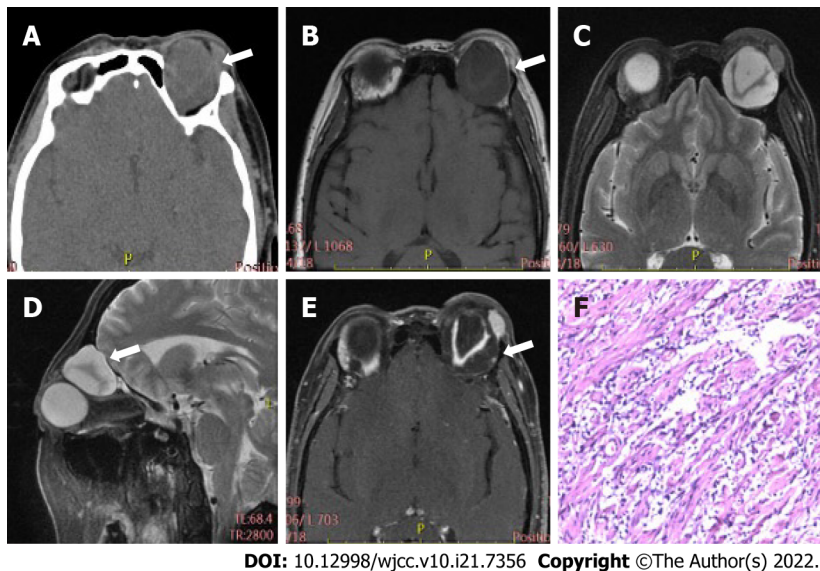
<sup>a</sup>*P* < 0.05.

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**Figure 1 Case 1, 71-year-old male, right orbital schwannoma.** A: On plain computed tomography (CT) scan, oval soft tissue nodule with smooth edge can be seen in the right orbit (indicated by the arrow), the size is about 25 mm × 16 mm × 15 mm, and the CT value is approximately 42 Hu; B-E: Magnetic resonance imaging showed that the focus was located in the extrapyramidal space of the muscle below the right orbit (indicated by the arrow), having slightly long T1, T2 signals, and the enhanced scan displayed uneven persistence moderate or obvious enhancement; F: Spindle cell can be seen microscopically.

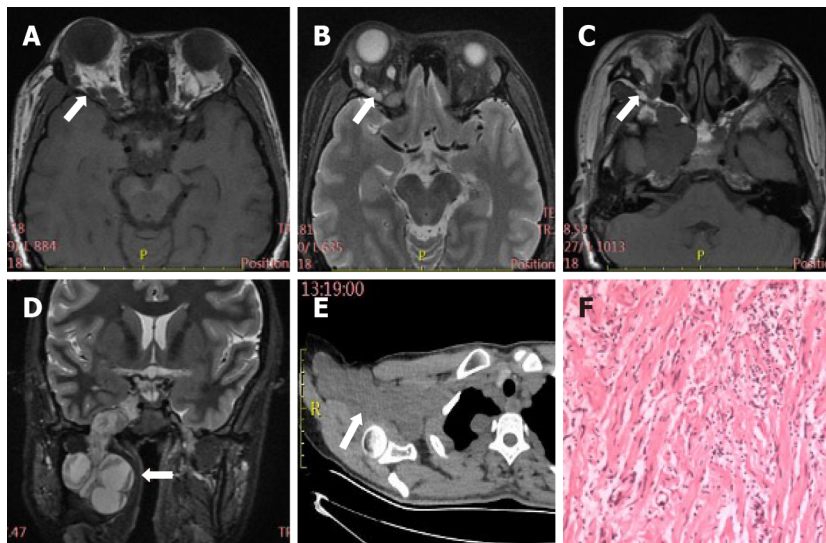
changes[18]. Based on cell morphology, schwannomas are divided into two types: Antoni A and B patterns. The Antoni A pattern often displays well-differentiated spindle cells with palisade nuclei, whereas the Antoni B pattern is characterized by bipolar and multipolar cells suspended in a loose myxoid matrix[19,20]. Imaging findings of schwannomas correlate with the biphasic pattern observed histopathologically. Most tumors dominated by Antoni A are solid, whereas those dominated by Antoni B are cystic and more vascular, which explains the diversity of imaging manifestations of neurilemmomas. MRI showed a slightly high signal in the Antoni A pattern and a significantly high signal in the Antoni B pattern on T2WI. On the contrast-enhanced scan, the enhanced signal of the Antoni B pattern was significantly stronger than that of the Antoni A pattern. Owing to cystic changes





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**Figure 2 Case 2, 53-year-old female, left orbital myxoid neurofibroma.** A: A non-uniform density mass of left orbit with a size of 33 mm × 30 mm × 21 mm was displayed by computed tomography (CT) (as shown by the arrow); the CT value was 21-60 Hu; B-E: Magnetic resonance imaging showed that the focus, which was mainly cystic (long T1 and T2 signals), was located in the extrapyramidal space above the left orbit (as shown by the arrow); "V" shaped nerve fibers were observed (equal T1 and T2 signals); on contrast-enhanced scan, the cystic part was not enhanced, while the solid part showed continuous and obvious enhancement; F: Microscopically, there is a large amount of mucus around nerve fiber cells.



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**Figure 3 Case 3, 37-year-old female, plexiform neurofibroma.** A and B: Magnetic resonance imaging plain scan showed nodules, with a diameter of 4-5 mm, had multiple slightly long T1 and long/slightly long T2 signals in the right orbit (indicated by the arrow); C and D: The focus grew into the brain through the supraorbital and inferior fissure (indicated by the arrow). An irregular mass, which was mainly cystic (long T1, T2 signals), with "V" shaped nerve fibers (equal T1 and T2 signals), could be seen in the right parasellar, pterygopalatine as well as infratemporal fossa; E: Plain chest computed tomography (CT) scan indicated multiple irregular and slightly low-density nodules, together with masses, located in the right brachial plexus distribution and the subpleural region, having an average CT value of approximately 36 Hu; F: Microscopically, nerve fiber cells with mucus around them could be seen.

and hemorrhage, schwannomas may have mixed signals. In a study by Young *et al*[21], the imaging characteristics of 13 patients with histologically proven schwannomas of the orbital frontal nerve were analyzed. Data showed extraconal location of all front nerve schwannomas, 10 of 13 patients had bone remodeling on CT, and 1 had calcification on pre-contrast CT. On pre-contrast CT, most lesions were heterogeneously isodense to hypodense. On post-contrast CT, all patients showed heterogeneous mild to moderate contrast enhancement. On T1-weighted MRI, most were heterogeneously iso- to hypointense, whereas on T2-weighted MRI, all were heterogeneously iso- to hyperintense, with portions of hypointensity within the tumor[21]. In comparison with our results, their data support an obviously high rate of bone involvement and a low rate of homogeneous density/signal intensity, which may be attributed to the single orbital nerve type.

Neurofibromas originate from non-myelinated, neoplastic Schwann cells, often presenting as smooth, solitary, and sometimes gelatinous masses with proliferation of collagen fibers and pathological mucinous matrix[22]. Because of mucoid degeneration, orbital neurofibromas often appear as uneven low-density masses on CT. On MRI, a characteristic target sign can be seen, the center of which is the involved thickened nerve bundle or dense collagen and fibrous tissue, showing equal or slightly longer T1 and shorter T2 signals, and the periphery is a mucus matrix with a hypersignal[23]. The tumor can extend to the intracranial region along the supraorbital fissure, and the surrounding orbital wall may have compressed changes if the tumor is large. Plexiform neurofibromas can be divided into superficial, tissue replacement, and invasive types, based on tumor location, range, and growth mode[24]. Superficial neurofibromas are limited to the skin and subcutaneous tissue without involution of the muscular layer, whereas the invasive type often shows diffuse invasive growth, involving more than three layers of skin structure[25]. The tissue replacement type is mostly nodular and grows along the nerve plexus with a clear boundary[26]. MRI scans of this type revealed tortuous and diffuse tumors along the nerve, multiple beaded, with a worm-like appearance, similar to case 3 in our study.

Schwannomas, neurofibromas, and plexiform neurofibromas are benign tumors with favorable prognoses[27]. Complete excision of these benign tumors with full effort to maintain capsular integrity is the mainstay treatment. However, for a patient whose documented serial imaging suggests a solitary schwannoma or neurofibroma affecting the apex, or where resection is not feasible, orbital decompression alone may be beneficial[10]. In our study, complete gross resection was performed for 33 patients based on imaging findings, and partial resection was performed for plexiform neurofibroma. The final diagnosis was clarified based on the pathology report of the postoperative specimens. These patients were followed-up for one-nine years, and there was no tumor recurrence.

The limitation of our study is the relatively small sample size, with only one case of plexiform neurofibroma. We believe that our results will encourage further studies using larger sample sizes and will help elucidate the imaging characteristics of all types of orbital PNSTs.

## CONCLUSION

PNSTs are rare neoplasms of the orbits. Different tumor types have distinct imaging characteristics. MR multi-direction imaging displays individual structures of the tumors, while CT can enable users to better observe the details of the involvement of the adjacent orbital bone. Since one imaging modality cannot see all the imaging variables, it is recommended to use plain and enhanced MRI scans as the main examination method, and CT scanning as an important supplementary means. A full understanding of the imaging characteristics of orbital PNSTs is helpful for improving diagnostic accuracy.

## ARTICLE HIGHLIGHTS

### Research background

Peripheral nerve sheath tumors (PNSTs) is a rare group of neoplasms in the orbit. Although computed tomography (CT) and magnetic resonance imaging (MRI) features could provide clues for the diagnosis of PNSTs, there are very few studies detailing the features of these tumors identified using imaging technology at present.

### Research motivation

The comprehensive characteristics of 34 patients with PNSTs were collected, and we found that imaging played as an important role in the diagnosis of this rare tumor.

### Research objectives

This study was designed to compare the clinical, pathological, CT, and MRI data in 34 patients with periorbital sheath tumors, including 21 schwannomas, 12 neurofibromas, and 1 plexiform neurofibroma.

### Research methods

All data were analyzed retrospectively in 34 patients with periorbital sheath tumors diagnosed using histopathology from January 2013 to August 2021.

### Research results

Schwannomas mostly occur in the intramuscular space with small tumor volume and rare bone involvement. Neurofibromas develop in the extrapyramidal space with larger tumor volume and more bone involvement. One case of plexiform neurofibroma showed tortuous and diffuse growth along the nerve, with a worm-like appearance on imaging.

## Research conclusions

Imaging manifestations have great value in the diagnosis of orbital peripheral nerve sheath tumors.

## Research perspectives

Further studies will elucidate the imaging characteristics of all types of orbital PNSTs using larger sample sizes, and focus on the value of imaging in the surgery of orbital PNSTs.

## FOOTNOTES

**Author contributions:** All authors have read and approved the manuscript; Dai M drafted part of the manuscript, and is responsible for the revision and publication fees; Wang T performed the literature review and drafted part of the manuscript; Wang JM, Fang LP and Zhao Y collected the patient data; Thakur A critically polished the language; Wang D interpreted the data.

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