Extradural Glomus Tumor of the Thoracic Spine: Case Report and Literature Review

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Research

Keywords: glomangioma, spinal mass, preoperative diagnosis

Posted Date: May 3rd, 2021

DOI: https://doi.org/10.21203/rs.3.rs-390250/v2

Abstract

Background: Glomus tumors are rare lesions that can arise anywhere along the spinal axis. Only thirteen cases have been reported. We report a case of a patient with a rare glomangioma of the thoracic spine. Following a review of the twelve previous cases is a discussion of a spinal glomus tumor with regard to clinical presentation, diagnosis, and treatment.

Methods: A single, recent case arising from the extradural space of the thoracic spine is described, followed by a literature review of spinal glomus tumors.

Results: Including our case, thirteen cases of a glomus tumor have been reported. The mean age at the time of diagnosis was 44.6 years (range: 22–73 years) and the male-to-female (M:F) ratio was 1.17:1. The lesion was located in the cervical spine in 1 patient, the thoracic spine in 7 patients, the lumbar spine in 4 patients, and the sacrum in 1 patient. All the tumors were benign; however, there was one report of local recurrence because of incomplete removal. There were no malignant transformations, metastases, or deaths reported with a mean follow-up of 20.09 ± 28.43 months (range: 2–90 months).

Conclusions: Spinal glomus tumor are rare, and the preoperative diagnosis is difficult. An overwhelming majority of glomus tumors are benign and are cured by simple local excision. Patients undergoing complete resection have an excellent long-term prognosis.

Trial registration: Not applicable.

Background

Spinal masses often present challenging diagnostic and therapeutic scenarios. A recent patient with an extradural mass in the thoracic spine, which was determined to be a glomangioma, illustrates this problem. This case led us to perform a literature review regarding this rare tumor, which we hoped would enable us to determine the tumor's imaging characteristics and biologic behavior, as well as the ability to establish criteria for a preoperative diagnosis. Our analysis of this patient and the current literature are presented.

Methods

A review of the patient's chart was conducted along with a review of the literature using a Medline search. Translations were performed whenever necessary. Details of the eight patients (including the current patient) are summarized in Table 1.

Case report

A 47-year-old man presented with a 5-month history of numbness below his umbilicus, especially in his left leg. The patient did not seek treatment until he developed an unsteady gait. Upon neurological examination, the patient had diminished sensation of pinprick and light touch below his umbilicus, and there was weakening of the myodynamics of both lower extremities. Interestingly, his reflexes were symmetric.

The hematologic examination was within normal limits. Magnetic resonance (MR) imaging showed an extradural lesion at the vertebral body of T7, extending to the T9 vertebral body, with low signal intensity on T1-weighted images (T1WIs); in addition, bright enhancement occurred following gadolinium administration (Figure 1). The lesion showed high signal intensity on T2-weighted images (T2WIs). The spinal cord was severely compressed, and the T8 neural foramen was enlarged. However, osseous destruction was not observed.

Surgical intervention was proposed because of the progression of the neurologic deficit. The patient underwent a T8-T9 hemilaminectomy and complete resection of the extradural tumor. The tumor had an irregular capsule, and slight bleeding occurred when it was incised. After undergoing posterior decompression, his sensory deficit was stable; however, he had regained normal strength in both legs and was living independently.

Histopathologic Examination

Permanent microscopic slides showed a hypercellular neoplasm with sheets of round-to-polygonal epithelioid cells. Individual nuclei were regular, round-to-ovoid, and lacked significant nuclear atypia or necrosis. The background stroma was hyalinized to a myxoid appearance. Scattered, dilated, capillary-sized vascular channels coursed through the lesion (Figure 2). The neoplastic cells were immune-negative for CD117, CK, CgA, Desmin, Factor , LCA, E-cadherin, NSE, Syn, and S-100. Immunoreactivity to SMA, vimentin, CD34, PAS, and CD99 (Figure 3) was noted. This staining pattern, in conjunction with the histological features, was consistent with the diagnosis of a glomangioma. This case is considered to be the second with these characteristics to be reported in the literature.

At his 3-month follow-up examination, the patient stated that he had fully recovered and had no difficulty in walking. He had returned to work and was very satisfied with the results of his surgery.

Results

A Medline search of the literature found 12 cases of a spinal glomus tumor. The details of those patients and our case are summarized in Table 1.

The mean age at the time of diagnosis was 44 .6 years (range: 22–73 years) and the male-to-female (M:F) ratio was 1.17:1.The lesion was located in the cervical spine in 1 patient, the thoracic spine in 7 patients, the lumbar spine in 4 patients, and the sacrum in 1 patient. All the patients underwent posterior decompression and one patient underwent emergency embolization of the segmental arteries (adhesive and coil embolization) because of profuse bleeding that could not be controlled with other measures. All 13 lesions were located in the extradural space of the spine. The dura was not involved in any case; however, bone structure involvement, erosion, and lytic lesions were found in 8 of 13 patients.

There were postoperative follow-ups with 11 of the 13 patients from the literature review, as well as our patient; the mean follow-up of the 13 patients was 20.09 ± 28.43 months (range: 2–90 months). The longest follow-up was 7.5 years. One case of local recurrence was found in the literature review. This was due to incomplete removal. No malignant transformations, metastases. or deaths occurred at the conclusion of the follow-up periods.

Discussion

Glomangiomas, a subset of glomus tumors that have a rich vascular network, are neoplasms of the glomus apparatus. Glomus tumors are benign neoplasms of the perivasculature. They arise from modified smooth muscle cells called glomus cells; they are located in the walls of the Sucquet-Hoyer canal, which is a specialized arterial-venous anastomosis central that controls blood flow for thermal regulation.

Glomus tumors are most commonly present in the subungual region of the finger; other common sites include the palm, wrist, forearm, and foot [1-4]. In fact, it is now recognized that the tumor also develops in sites where a normal glomus body may be sparse or even absent, such as the patella, long bones, stomach, colon, liver, nerves, eyelids, nose, mediastinum, and mesentery [5-12]. A subungual presentation is more common in women; however, no sex predilection is evident at other loci. The male-to-female (M:F) ratio was 1.17:1 in our series. Multiple tumors are more often hereditary and painless; they are more frequent on the arms and have no sex predilection. Multiple lesions may rarely be associated with arteriovenous fistulae, nodular lesions of finger joints, type II multiple endocrine neoplasia, and bone changes such as brachydactyly [13-15].

Glomus tumors are composed of varying proportions of glomus cells, vascular structures, and smooth muscle cells. Depending on the predominant component, there are three variants of glomus tumors. When glomus tumors have prominent glomus cells, poor vasculature, and a scant smooth muscle component, they are classified as solid glomus tumor. Whereas glomus tumors with a prominent vascular component, are classified as glomangiomas. Finally, when there is a prominent smooth muscle component, the tumors are classified as glomangiomyomas. A solid glomus tumor is the most common variant (75%),

followed by glomangioma (20%) and glomangiomyoma (5%) [16]. In our series, 2 of 13 cases were deemed to be glomangiomas; the other 11 were solid glomus tumors [17-28].

Microscopically, glomus tumors present as uniformly round-to-ovoid glomus cells arranged as lobules, strands, or broad sheets in a brickwork-like manner. The association with vascular structures includes indistinct capillaries in the walls of the surrounding large blood vessels as well as highly altered vessels. Uniform epithelioid glomus cells with inconspicuous-to-well-defined nucleoli are found between the sinusoidal vessels. Despite their epithelioid appearance and intimate association with vasculature, glomus cells are immunoreactive with vimentin and SMA. However, a small number of these tumors may coexpress myogenic markers and CD34. The current case was immunoreactive with SMA, Vimentin, CD34, PAS, and CD99.

Irrespective of their anatomical location, glomus tumors are usually benign; however, rare malignant cases have been described [29, 30]. Recently, Folpe et al. [31] proposed a classification of atypical and malignant glomus tumors. Malignant glomus tumors are defined as tumors with: (1) large size (> 2 cm) and deep location, (2) atypical mitotic figures, or (3) nuclear atypia. In contrast, glomus tumors of uncertain malignant potential are superficial in location and show a high mitotic activity, are large in size, or are deeply located. However, in their series of unusual glomus tumors, Folpe et al. [31] defined the tumor location as either superficial to the peripheral soft tissue or deep to it. In their study, there were 9 deeply located peripheral soft tissue glomus tumors, and all of them were larger than 2 cm; however, only 5 cases metastasized. In a series of 32 gastrointestinal glomus tumors, which by definition were deeply located, only 1 case metastasized, although 19 were larger than 2 cm [32]. In our case review, all of the lesions were > 2cm, and deeply located. The mean follow-up of 13 patients together with the current patient was 20.09 ± 28.43 months (range: 2–90 months); the longest follow-up was 7.5 years. No malignancies had appeared by the conclusion of the follow-up periods. Therefore, the risk of metastases of the deep-seated glomus tumors to the peripheral soft tissue appears to be markedly different from those located in other deep regions. An extracutaneous, deep location should not be considered to be a sign of malignancy. On the basis of these criteria and the lack of atypical mitotic figures, our patient's tumor would be classified as benign. Occasionally, glomus tumors occur in a hereditary manner. The gene has been localized to chromosome 1p21–1p22, which codes for glomulin, a protein thought to play a role in vascularization [33, 34].

The preoperative diagnosis of spinal glomus tumor is very difficult. Usually, the plain radiograph is unremarkable for a spinal glomus tumor. Most patients with a spinal mass undergo a CT scan and magnetic resonance imaging (MRI). Although CT images can show well-defined lesions in a few cases, MR images are considered to be an excellent diagnostic modality. On T1-weighted MR images, glomus tumors may present as a sharply marginated lesion with hypointensity or isointensity, while on T2WIs they are mostly hyperintense. Furthermore, they usually show strong intense enhancement on T1WIs after intravenous administration of gadolinium-DTPA [4]. Most glomus tumors are surrounded by an envelope. T2WI and T1WI after intravenous administration of contrast medium can detect the envelope as a low signal intensity rim. The envelope, which may be incomplete, may be the consequence of a secondary reaction with the surrounding tissue. An envelope-like structure was observed in our case. The MR images of our case are fairly similar to the MR features of the other seven cases. However, a characteristic finding in our case was signal-void fistular lesion in the tumor. The features of the fistular lesion were similar to the images of vascular structures. It also was comparable to the histological findings of tissue with a prominent vascular component. The fistular lesion, due to the direction of flow, was identified on the sagittal images but not on the axial images.

In our case, the differential diagnosis included primary extradural masses or intradural, extramedullary lesions of the spine. Schwannomas, which are the most common extradural tumors, show the typical dumbbell-shaped appearance (extra- and intra-dural portions) in 50% of cases; however, 50% are completely extradural. Schwannomas on MRI images tend to have low signal intensity on T1WI and have brightly increased signal intensity on T2WI due to the high-water content of these lesions. These lesions usually were markedly enhanced following gadolinium administration. The MR findings of the eight cases had similar MR features of the schwannomas, except for the signal-void fistular lesion. Ganglioneuromas and neuroblastomas are tumors of childhood, which often appear with large paraspinal tumor masses.

Choice of therapeutic strategy is predicated according to lesion location and clinical presentation. An overwhelming majority of glomus tumors are benign and are cured by simple local excision. The postsurgical recurrence rate is approximately 10%,

because of incomplete removal. In the literature we reviewed, with the longest follow-up being 7.5 years, there was one report of local recurrence because of incomplete removal; however, no malignant transformations, metastases, or deaths occurred by the conclusion of the follow-up period.

Conclusions

In summary, spinal glomus tumor, although rare, deserve consideration during the differential diagnosis of spinal lesions. Preoperative diagnosis is difficult; however, certain imaging features can give more information to base this diagnosis on, and raise its level of certainty. Differentiation of glomus tumor from schwannomas can be accomplished on the basis of histology and immunostaining. An overwhelming majority of glomus tumors are benign and are cured by simple local excision.

Declarations

Ethics approval and consent to participate

Written consent was obtained from the study participants patient.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests.

Funding

This work is supported by grants from the Liaoning Science and Technology Plan Project(No.2018225094).

Authors' contributions

ZJH did the conceptual design, experimental planning and realization, editing and reviewing of this study and manuscript. LXX managed and organized the study, while also helping with experimental planning and realization. All authors read and approved the final manuscript for publication.

Acknowledgements

Not applicable

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Table

Table 1
Summary of spinal glomus tumor in the literature

	Year	Gen-der	Age (yrs)	Presenting symptoms	Location	Treatment	Size (cm)	Biologic behavior	Pathology	Follow-up (months)
ni[27]	1990	F	22	Sacral pain	Sacrum	Sacral laminectomy	N/A	Benign	Solid glomus tumor	N/A
5]	1991	M	49	Back pain, compressive myelopathy	T2 pedicle vertebral body	T2-3 costotrans versectomy	>2	Benign	Solid glomus tumor	90
[24]	1996	F	45	Back pain	L1 pedicle	Posterior laminectomy and resection	>2	Benign	Solid glomus tumor	2
]	2002	F	55	Back pain compressive myelopathy	T4 vertebral body	Transthoracic T4 corpectomy	5	Benign	Solid glomus tumor	3
]	2002	F	37	Unsteady gait	C7-T2 extradural space	C7-T2 laminectomy and partial resection of tumor	>4	Benign	Glomangiomas	6
17]	2005	M	50	Chronic lumbago	L1-L2 extradural space	Hemilaminectomy of L1-L2	3	Benign	Solid glomus tumor	12
dis[19]	2007	M	44	Radicular symptoms	L3 vertebral body, extradural space at L3-L4, L3 neural foramen,abdominal cavity	Posterior decompression, angiographic embolization, Transthoracic resection,360 degree recontruction and fusion	12	Benign	Solid glomus tumor	6
	2015	F	45	neurological symptoms	upper thoracic vertebra of T2-T4	posterior decompression tumor resection corpectomy	5x6	Benign	Solid glomus tumor	60
	2009	M	39	chronic lumbago	T12–L1	tumor resection corpectomy	4x5	Benign	Solid glomus tumor	15
	2017	M	26	progressive paraparesis paresthesia	T1-T2	prone position with intraoperative C-arm guidance tumor resection	N/A	Benign	Solid glomus tumor	N/A
	2019	M	48	chest discomfort intermittent back pain	T2–T4	Tumor resection unilateral fusion with pedicle screws at the T2 to T4 level	5.6x3,8	Benign	Solid glomus tumor	18
]	2012	F	73	upper and lower back pain	T11	percutaneous CT-guided radiofrequency ablation (RFA)	1.5x0.6	glomus tumor of uncertain malignant potential	Solid glomus tumor	6
article	2020	M	47	Compressive myelopathy	T7-T9 extradural space	T8-T9 hemilaminectomy	3	Benign	Glomangiomas	3

N/A:not available

Figures

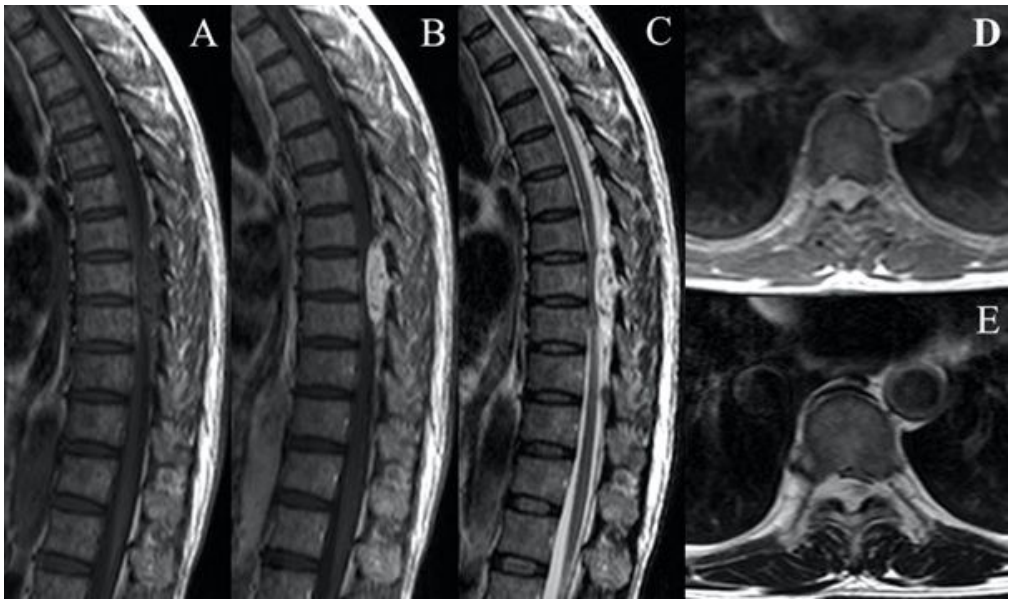

Figure 1

Magnetic resonance imaging showing an extradural lesion in the vertebral body of T7, extending to the T9 vertebral body, compressing the thecal sac. A: The mass is of low signal intensity on sagittal T1-weighted images; B: Following gadolinium administration, sagittal T1-weighted images showing brightly enhancement; C: Abnormal high signal intensity on sagittal T2-weighted images was limited by the tumor envelope. There were numerous signal-void fistular lesions on sagittal T1WI, T2WI and postcontrast T1WI in the lesion (A, B, C). D: Enhanced axial T1-weighted images showing that the T8 neural foramen was enlarged; E: An axial T2-weighted images showing that the T8 neural foramen was enlarged.

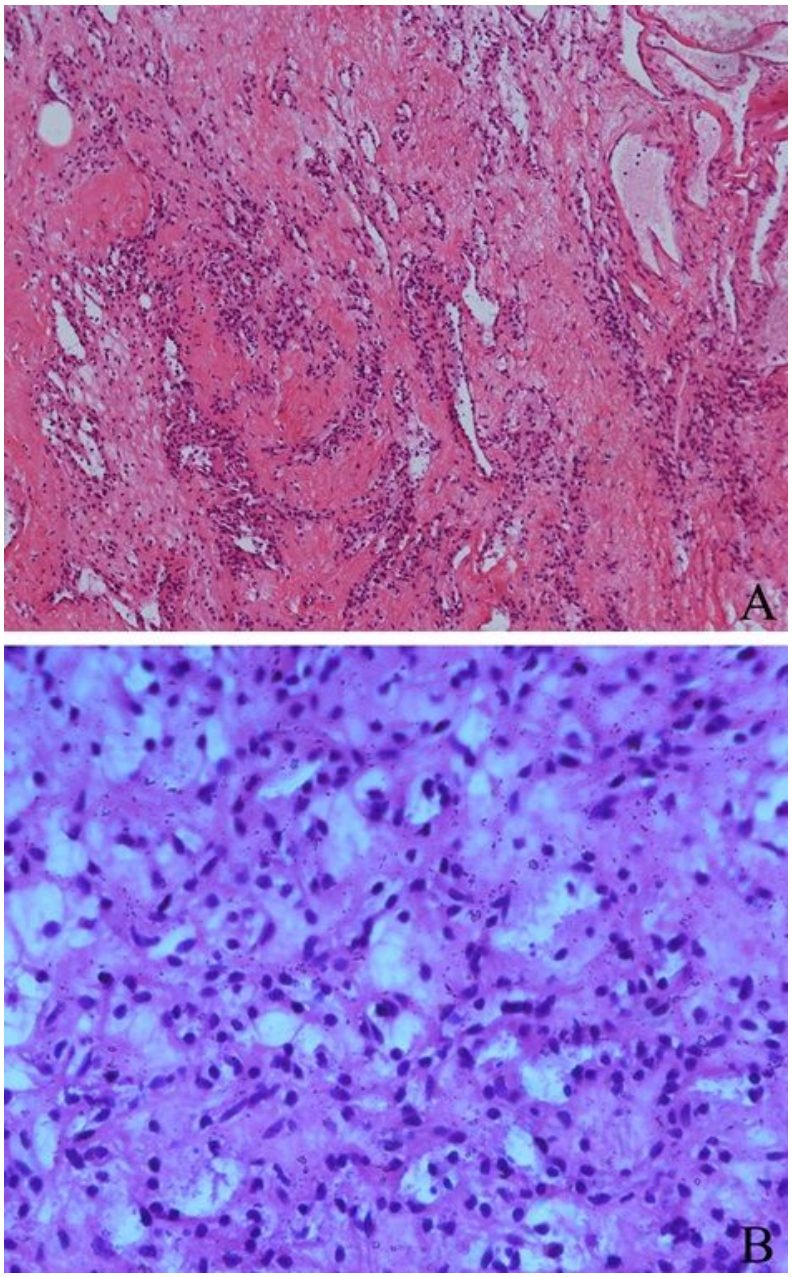

Figure 2

A: The neoplasm is composed of numerous, small-to-medium branched vessels surrounded by small, round-to-oval uniform cells (H&E; original magnification ×100); B: The neoplastic cells have sharply outlined round-to-oval nuclei with bland chromatin and scant-to-moderate eosinophilic cytoplasm (H&E; original magnification ×400).

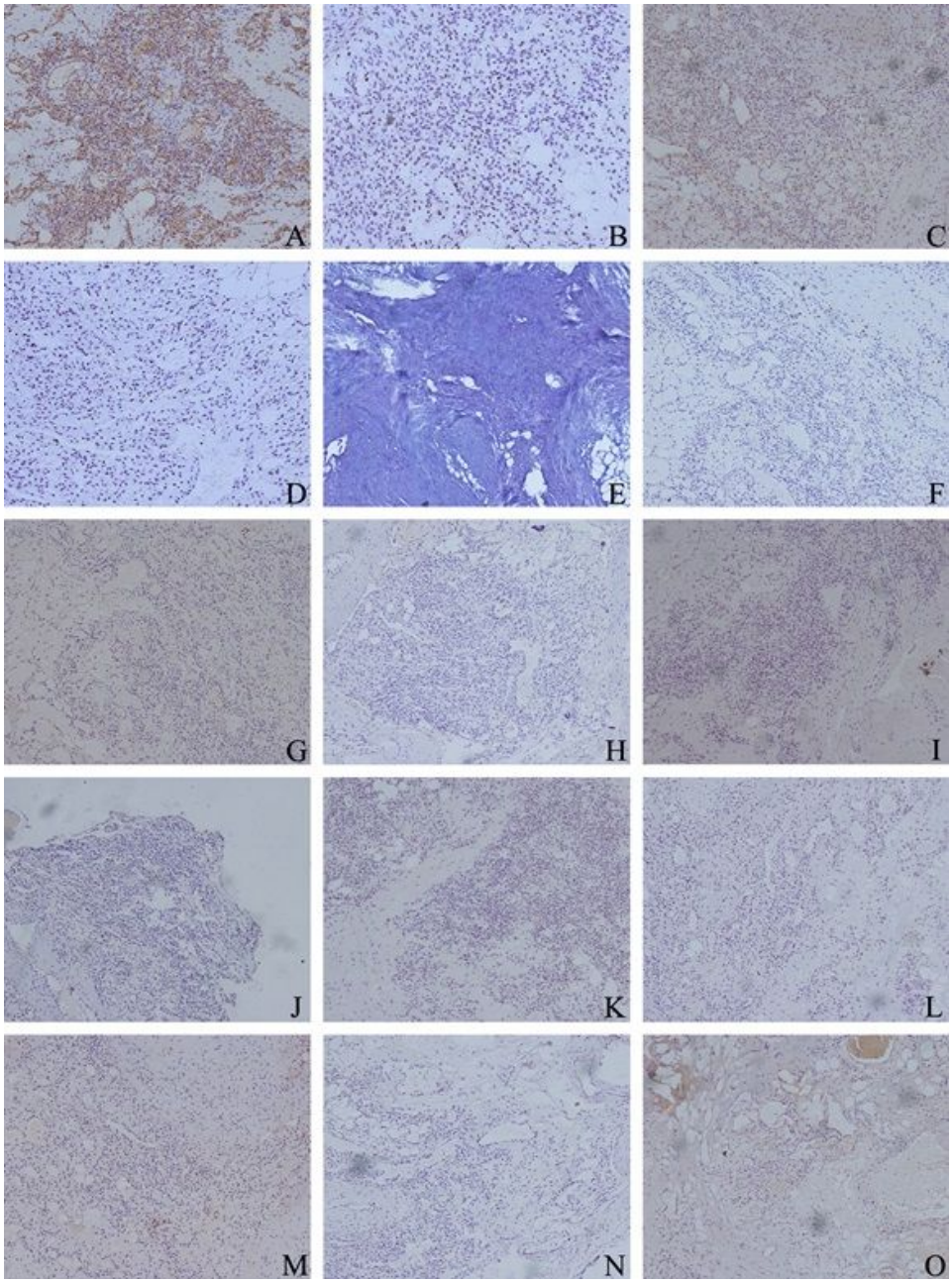

Figure 3

Immunohistochemically, tumor cells stained strongly for CD34, vimentin, and SMA (Figures 3A-3C), and a few cells stained faintly for CD99 and PAS (Figures 3D and 3E). Tumor cells were negative for CD117, CK, CgA, Desmin, Factor Ⅷ, LCA, E-cadherin, NSE, Syn, and S-100 (Fig. 3F-3O). A: CD34, original magnification ×100; B: Vimentin, original magnification ×200;C: SMA, original magnification ×100; D: CD99, original magnification ×200; E: PAS, original magnification ×100; F: CD117, original magnification ×100; G: CK, original magnification ×100; H: CgA, original magnification ×100; I: Desmin, original magnification ×100; J: Factor Ⅷ, original magnification ×100; K: LCA, original magnification ×100; L: E-cadherin, original magnification ×100; M: NSE, original magnification ×100; N: Syn, original magnification ×100; O: S-100, original magnification ×100.