J Neurosurg Spine 7:248–253, 2007

Pigmented villonodular synovitis associated with pathological fracture of the odontoid and atlantoaxial instability

Case report and review of the literature

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✓Pigmented villonodular synovitis (PVNS) is a proliferative disorder of the synovium with a predisposition for the appendicular skeleton. Rarely PVNS can arise from the spine, where this disorder usually presents with localized or radicular pain secondary to involvement of the posterior elements. The authors report the case of an 82-year-old woman who presented with long-standing neck pain and acute upper-extremity numbness and weakness. Computed tomography imaging revealed a mixed sclerotic and lucent lesion affecting the dens and right lateral mass of C-2. There was also a pathological fracture at the base of the dens with 8 mm of anterior dens displacement. Magnetic resonance imaging demonstrated a diffusely infiltrative process that was nonenhancing. Because of instability, the patient underwent transarticular screw fixation, and a biopsy of the lesion was also performed at this time. Histopathological analysis was consistent with a diagnosis of PVNS. To the authors' knowledge, this is the first report of PVNS involving the C-2 vertebra or causing a pathological fracture. *(DOI: 10.3171/SPI-07/08/248)*

KEY WORDS • **atlantoaxial instability** • **odontoid** • **pigmented villonodular synovitis**

PIGMENTED villonodular synovitis is a rare lesion of unknown cause arising from the synovium of the joint, tendon sheath, or bursa. Although the lesion is benign, it can be locally aggressive and is associated with a significant rate of recurrence after resection.[22]

Most commonly seen in the joints of the appendicular skeleton, PVNS affects the knee in up to 80% of cases but also commonly affects the hip, ankle, elbow, and digits.[22,26] This lesion rarely presents in the spine, with only 42 cases reported in the literature to our knowledge (Table 1).[4–7,10–12,16–19,21,24,25,28,29] In the spine, the disease has a predilection for the posterior elements, and most patients present with complaints of either localized or radicular pain.[10] Treatment has centered on resection, with gross-total resection being the goal if possible.[10,12]

We describe the case of an 82-year-old woman who was found to have PVNS of the atlantoaxial articulation with infiltration of the dens and a pathological odontoid fracture after presenting with neck pain and progressive quadriparesis. This is the first reported case of PVNS affecting this region of the spine and the first reported case of PVNS causing a pathological spinal fracture.

Case Report

Presentation and Examination. This 82-year-old woman had complained of high cervical neck pain for about 1 year and presented to the emergency department 5 days after experiencing an acute onset of increased neck pain associated with bilateral hand numbness and quadriparesis. Her

Abbreviations used in this paper: CT = computed tomography; PVNS = pigmented villonodular synovitis.

history was significant only for hypertension, and she had been very active and living independently before this event. She denied any history of trauma.

On initial examination, the patient had Grade 5−/5 strength in grip bilaterally and Grade 4/5 strength in the triceps bilaterally. She had Grade 5/5 strength in all other muscle groups, although she did have some unsteadiness of gait. Her sensation was intact to all modalities, and her reflexes were symmetrical and nonpathological.

Operation. Computed tomography scanning of the spine revealed a mixed sclerotic and lucent lesion involving the odontoid process and right lateral mass. A pathological fracture through the base of the dens was noted, with the dens displaced 8 mm anterior to the body of C-2 (Fig. 1). Magnetic resonance imaging revealed a diffusely infiltrative process affecting the body of C-2 and the dens. The lesion was nonenhancing and heterogeneously hyperintense on T2-weighted magnetic resonance images and short tau inversion recovery sequences (Fig. 2).

Because of the instability, the patient underwent surgery to stabilize the region with transarticular screws and a posterior Dickman–Sonntag construct using allograft. During the procedure, the lytic lesion was noted to involve the superior surface of the right pars interarticularis. Analysis of a biopsy sample revealed synovial-type tissue with a fibrohistiocytic reaction, multinucleated giant cells, and hemosiderin deposits consistent with PVNS (Fig. 3).

Postoperative Course. The patient tolerated surgery well and has made a complete recovery. On consultation, she has elected to have the lesion monitored with serial imaging and to forgo any attempt at obtaining a gross-total resection at this time. She has undergone monitoring with biannual CT scans, which have not shown evidence of recurrence at the 2-year follow-up (Fig. 4).

Discussion

Pigmented villonodular synovitis is a rare proliferative disorder of the synovium that is most commonly seen in the appendicular skeleton where it causes symptoms of joint pain, swelling, and stiffness. Its occurrence in the spine was first described by Kleinman et al.[18] in 1980, and 41 additional cases have since been reported to our knowledge.

Although the lesion of PVNS has been assigned a variety of names, including nodular tenosynovitis, fibrous histiocytoma of the synovium, giant cell synovioma, and fibrous xanthoma of synovium,[11] all of these terms describe a similar lesion, of which two primary forms, localized and diffuse, are recognized. The localized form of PVNS affects only a portion of the synovium and tends to affect the small joints of the hands and feet.[23,24,28] It is often termed "giant cell tumor of the tendon sheath" when seen to arise from this anatomical structure.[28] The diffuse variant affects the entire synovial lining of the joint and tends to affect the larger joints such as the knee and hip.[23] The diffuse type tends to invade surrounding tissue, may cause the erosion of bone or other neighboring structures, and has a greater propensity for recurrence than the localized type.[11,30]

When the spine is involved, the disease typically arises from the posterior elements because the facet joints contain synovium. One unique aspect of the case we present is that the PVNS does not appear to arise from the posterior elements. This distinctive feature can be explained by the fact that the atlantodental joint also contains a sheath of synovium, and therefore is another region of the spine from which PVNS may originate outside of the facet joints.

The cause of PVNS is a subject of speculation, and various theories on the origin of the disease, including neoplasia, hyperplasia, inflammatory reaction, metabolic derangement, and recurrent hemorrhages, have been proposed.[11,13,29] Several authors have suggested a traumatic cause, citing a history of recent trauma in several patients with the lesions.[2,9,14,16] This has not been a universal observation, however, and there is no statistical evidence to support this link.[12] Additionally, experimental attempts to reproduce this lesion in animal models have failed.[31] Recent evidence, including evidence of clonality, high rates of local recurrence after partial resection, and rare examples of malignancy indicate that the lesions are more likely neoplastic than reactive in nature.[1,3,8,20]

Pathological examination of PVNS reveals a lesion composed predominantly of closely aggregated round to polygonal mononuclear epithelioid cells with varying numbers of multinucleated giant cells, lymphocytes, siderophages, and xanthoma cells. There are varying amounts of fibrous tissue, and the overall architecture may be nodular or villiform. Mitoses are frequently observed, as are hemosiderin deposits, although the degree to which an individual lesion will manifest these characteristics is variable.

Pigmented villonodular synovitis of the spine appears to affect both sexes and patients of all ages equally, with a mean age of 38 years and cases reported in patients ranging in age from 13 to 84 years.[10] It most frequently affects the cervical spine (52% of cases), followed by the lumbar (29%) and thoracic (17%) spine.[10] The most common symptom prompting workup is localized pain, although radiculopathy with associated pain and motor weakness is not uncommon.

Imaging demonstrates characteristic but nonspecific findings, making preoperative diagnosis difficult, especially considering the rarity of the lesion. Pigmented villonodular synovitis preferentially affects the posterior elements of the spine, a finding reported in the large majority of reported cases (Table 1, Fig. 5). For instance, facet joint involvement was documented in 10 of 11 cases in one series, and a facet joint origin was suggested in most of these.[10] In three cases, however, the anterior structures of the spine appeared to be primarily affected.[12,24] Although the origin of the lesion could not be definitively identified in any of these cases, Pulitzer and Reed[24] speculated that it may have been the synovial membranes of the accessory joints of the vertebral column. Another possibility would be an anterior growth pattern from the posterior elements with only a small, possibly overlooked connection. Computed tomography scanning demonstrates evidence of bone injury and epidural expansion of the lesion in at least 70% of cases.[10] The lesion may appear to be hyperattenuated because of the presence of hemosiderin.[28] Magnetic resonance imaging evaluation better delineates the soft-tissue mass, destruction of surrounding structures, and compression of neural elements.[12,15] On T1-weighted images a heterogeneous lesion of intermediate to low signal intensity is revealed, whereas T2-weighted sequences reveal a hypointense lesion secondary to hemosiderin deposition,[15] a common but not universal finding.[23] Administration of Gd

TABLE 1

*Summary of previously reported patients with PVNS**

Authors & Year	Age, (yrs) Sex	Location	Treatment	Follow-Up	Symptoms	Comments
Kleinman et al., 1980	65, F	C3–6 facet	biopsy	stable at 8 mos	neck pain, myelopathy	
Campbell & Wells, 1982	54, F	L4–5 facet	GTR	NED at 31 mos	back & radicular pain	
Pultizer & Reed, 1984	35, F	C1–4 paravertebral	GTR	NED at 11 yrs	hypopharyngeal mass/cough	speculated to arise from AJS
	23, M	L5–S1 paravertebral/ pars pedicle	en bloc resect	NA	incidental	speculated to arise from AJS
Weidner et al., 1986	48, F	L5–6 facet	1) GTR; 2) GTR; 3) radical resect	NED at 8 yrs	back pain	
	34, F	L4–5 facet	incomplete resect	NED at 44 mos	back pain, sensorimotor radiculopathy	
Retrum et al., 1987	81, F	L5–S1	GTR	NA	back pain	
Karnezis et al., 1990	37, F	C6–7 posterior elements	GTR	NED at 79 mos	neck pain	
Khoury et al., 1991	61, F	L4–5 facet	GTR	NA	radiculopathy	
	84, M	C4–5 facet	GTR	NA	SCC/ myelopathy	
Kuwabara et al., 1992	25, F	T8–11 VB/paravertebral	1) incomplete resect w/ XRT & chemo; 2) GTR	NED at 7 yrs	SCC	
Mahmood et al., 1992	43, F	C6–7 facet	GTR	NA	incidental	
Titelbaum et al., 1992	51, F	L4–5 facet	GTR	NA	back & radicular pain	
Clark et al., 1993	23, M	T7–8 facet	GTR	NED at 42 mos	SCC/paraparesis	
Gezen et al., 1996	19, M	T10–11 posterior elements/pedicle/VB	incomplete resect	stable at 16 mos	back pain & SCC w/paraparesis	
Giannini et al., 1996	21, F	T-3 facet	GTR	NED at 53 mos	neck or back pain in 9/12, radiculopathy in 6/12, weakness in 1/12 (patient of Clark et al., included in analysis)	
	26, M	C-6 facet	GTR	NED at 11 mos		
	29, M	L5–S1 facet	1) GTR; 2) radical resect	NED at 23 mos		
	37, F	C-7 facet	GTR	NED at 66 mos		
	37, F	C-7 facet	GTR	NED at 9 yrs		
	38, M	C4–5 facet	1) GTR; 2) GTR	NED at 55 mos		
	40, M	T-11 ?	GTR	NED at 64 mos		
	42, M	L-3 ?	incomplete resect	stable at 14 mos		
	43, M	C-5 facet	GTR	NED at 36 mos		
	44, M	C5–6 disc	1) biopsy; 2) GTR	NED at 6 mos		arose from the pst disc w/o osseous or facet involvement
	67, M	L-3 facet	NA	NA		
Bruecks et al., 2001	13, F	T7–8 posterior elements & pedicle	GTR	NA	back pain & SCC w/paraparesis	
Furlong et al., 2003	17, M	sacrococcygeal	GTR	LTF	localized spine pain in 12 patients; radicular pain, hyperactive reflexes, or weakness present in 10	pst element/facet involvement in ing 11/11 w/ imaging
	21, F	T4–5	GTR	LTF		
	23, M	C2–3	1) incomplete resect 2) GTR; 3) XRT	residual tumor at 5 yrs		
	25, M	C3–5	GTR	LTF		
	25, F	L4–5	1) incomplete resect 2) GTR	residual tumor		
	29, F	L4–5	en bloc resect	NED at 4 yrs		
	31, M	C5–6	GTR	NED at 9 yrs		
	32, F	C3–4	GTR	NED at 3 yrs		

(continued)

TABLE 1

Summary of previously reported patients with PVNS (continued)*

32. M	C3–4	1) incomplete resect 2) 2nd resect & XRT	stable at 2 yrs
35. M	cervical spine	GTR & XRT	NED at 7 yrs
39. F	cervical spine	biopsy	NED at 10 yrs
39. F	C5–6	GTR	LTF
43. F	C5–6	incomplete resect	LTF
44. M	cervical spine	GTR & XRT	NED at 4 mos
44. F	C4–5	biopsy	LTF

* AJS = accessory joint synovium; chemo = chemotherapy; GTR = gross-total resection; LTF = lost to follow-up; NA = not available; NED = no evidence of disease; pst = posterior; SCC = spinal cord compression; VB = vertebral body; resect = resection; XRT = radiotherapy; ? = exact level is unknown.

reveals avid enhancement in most,[23] but not all, cases.[11] The differential diagnosis, based on imaging findings, includes osteoblastoma, giant cell tumor of bone, aneurysmal bone cyst, and reactive synovial proliferation.[10,23]

Treatment of PVNS is surgical with gross-total resection being the goal. Giannini et al.[12] calculated a recurrence rate of 18% after gross-total resection in their review, which is similar to that reported for peripherally manifesting lesions.[27] Furlong et al.[10] reported no recurrence in five of five patients who underwent gross-total resection and recurrence in all four patients who underwent a subtotal resection. Although the small number of cases reported precludes meaningful statistical conclusions, the authors thought that recurrence was correlated with diffuse growth pattern, diminished osteoclast-like giant cell component, epidural involvement and soft-tissue extension beyond the bone margins, and patient age younger than 30 years.[10] Interestingly, factors such as tumor size and mitotic activity did not correlate with recurrence.[10] Whatever the cause, recurrences appear to be best treated with additional resection, which has been shown to provide disease control even after multiple recurrences.[29] In those cases in which a gross-total resection is not possible, subtotal resection may often provide disease control.[12] This has been the finding in our case in which the patient has remained stable for 2 years after biopsy only. Follow-up monitoring should be meticulous, however, and reoperation with an aim toward gross-total resection should be considered with clinical or imaging-documented progression. We are currently monitoring our patient with biannual CT scans. With continued stabil-

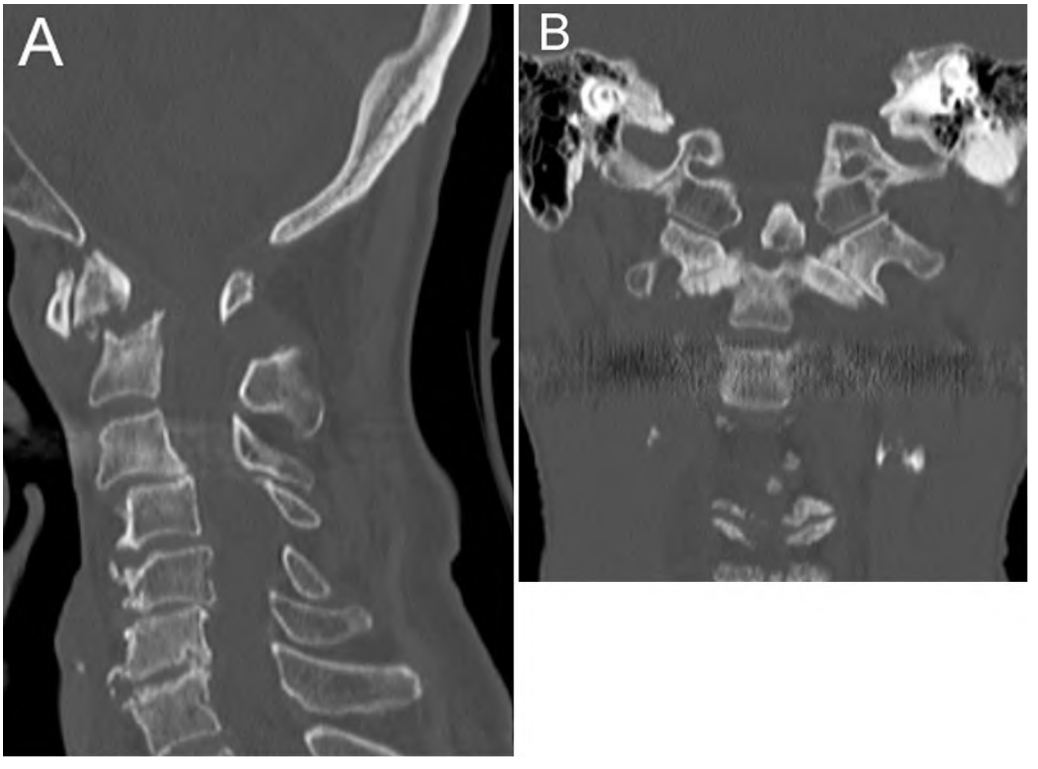

FIG. 1. Sagittal (A) and coronal (B) CT reconstructions showing a mixed sclerotic and lucent lesion involving the atlas and a pathological fracture through the base of the dens.

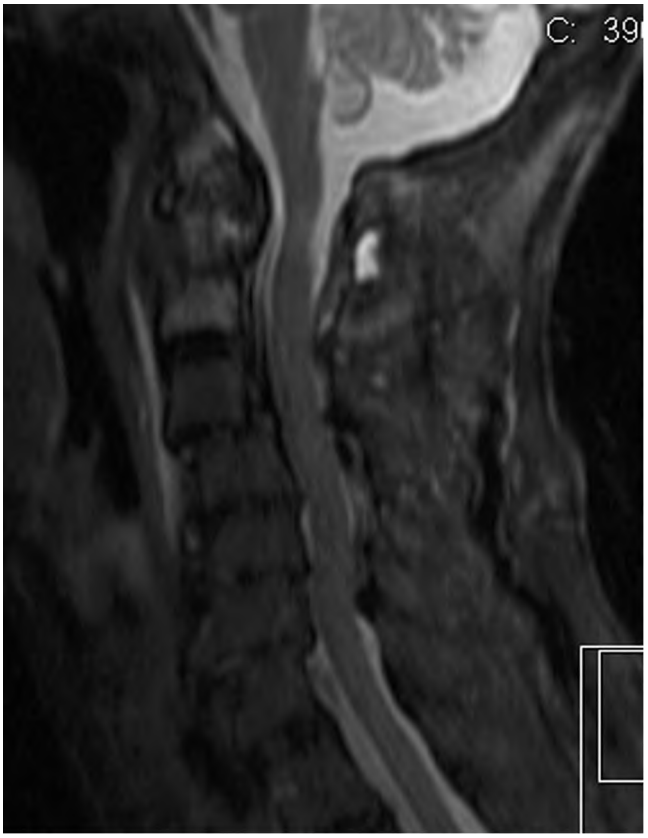

FIG. 2. Sagittal T2-weighted magnetic resonance image demonstrating a heterogeneously hyperintense lesion infiltrating the atlas and dens.

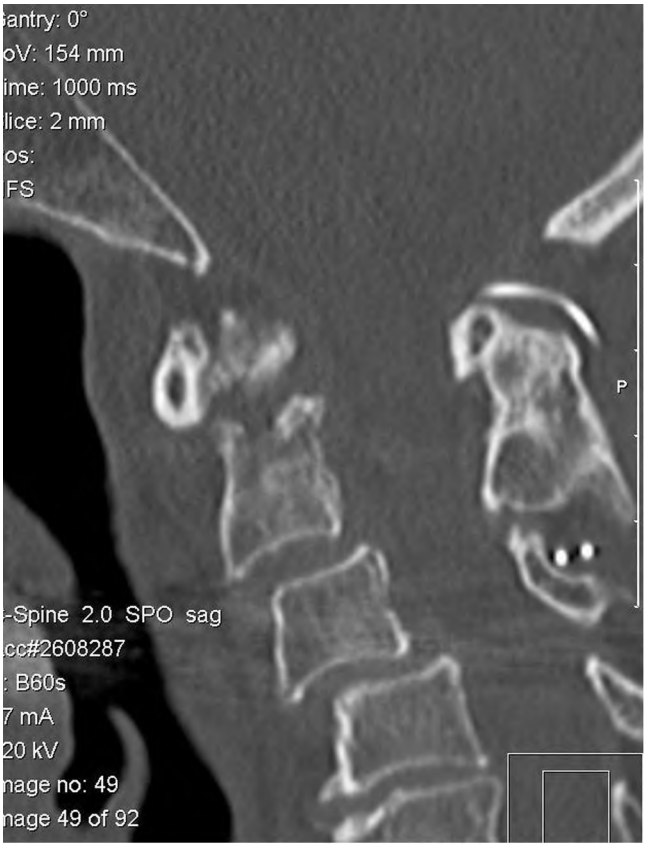

FIG. 4. A CT scan obtained 2 years postoperatively showing solid bone fusion and lack of disease progression.

ity, we may decrease the frequency of imaging but will continue to monitor her for the long term, as there is no clearly defined time period after which progression is no longer noted. Although there have been several reports on the use of radiotherapy in the treatment of this disorder, its efficacy remains unproven.[7,10,12] Its use is, however, associ-

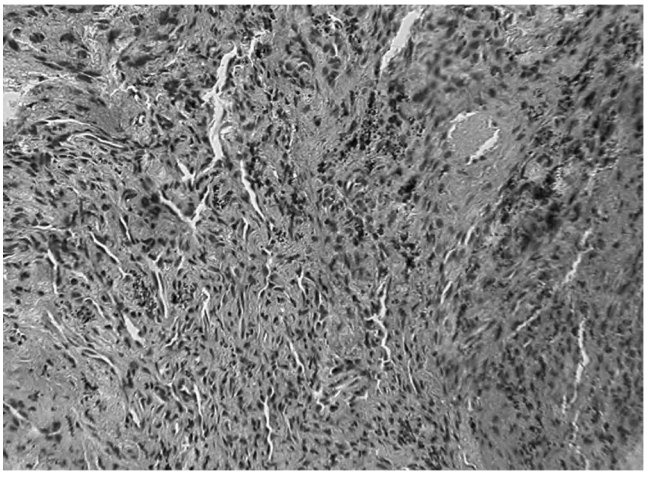

FIG. 3. Photomicrograph revealing synovial-type tissue with a fibrohistiocytic reaction, multinucleated giant cells, and hemosiderin deposition. Original magnification × 20.

	Anterior	Posterior	Unspecified	**Total**
Cervical	2	9	11	**22**
Thoracic	1	4	2	**7**
Lumbar	0	8	4	**12**
Sacral	0	0	1	**1**
Total	**3**	**21**	**18**	**42**

FIG. 5. Representation showing the location of spinal involvement of the 42 cases previously presented in the literature.

ated with wound breakdown and a small risk of radiation-induced sarcoma, and it should probably be reserved for cases of unresectable progressive disease.

Conclusions

We present an unusual case of PVNS affecting the atlantoaxial articulation. We believe it is the first reported case of this lesion affecting this joint as well as the first case associated with a pathological fracture of the spine. The lesion has remained stable for 2 years after posterior fixation and biopsy alone, underscoring the benign nature of the disorder. Additional resection and radiotherapy are options in cases of recurrence.

Acknowledgment

We thank Kristin Kraus for her assistance in the editorial preparation of this paper.

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Manuscript submitted February 6, 2007.
Accepted April 10, 2007.

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