

Case Report

# Severe Nightly Thoracic Pain Presenting during Pregnancy: A Case Report

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## Keywords

Pregnancy · Ependymoma · Pain

## Abstract

We present the case of a woman who developed severe nightly thoracic pain during pregnancy without neurological deficits upon examination. Spontaneously after childbirth, the pain was markedly reduced. Further investigation showed that her pain was caused by an ependymoma in the cervicothoracic spinal cord. Gross total resection was accomplished, and the patient has been free of pain ever since. With this case, we want to draw attention to a rare, but possibly very disabling, cause of increasing nightly thoracic pain during pregnancy. Spontaneous improvement after childbirth could erroneously cause postponement of further investigation.

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## Introduction

Ependymoma is a rare primary tumor of the central nervous system and can present anywhere along the ventricular system and spinal cord. Surgical intervention is usually necessary to ensure favorable neurological outcome [1, 2]. It is known that specific types of central nervous system tumors (for instance meningiomas) can accelerate in growth during pregnancy [3].

A spontaneous improvement of pain after childbirth could erroneously cause postponement of further investigation.

### Case Report

We describe the case of a previously healthy, 31-year-old woman, who presented in the last trimester of her pregnancy with pain between the shoulder blades. She was admitted to the obstetrics department because of this pain, and the neurology department was consulted. The pain had started in the sixth month of her pregnancy with her first child. The pain arose each night 2 h after she went to sleep, and it was described as unbearable. Our patient would wake up every night because of the pain. The pain was not correlated with breathing. There was no pain radiating to her arms or legs. At the first neurological examination, no abnormalities could be found, and no final diagnosis was made. Muscle and joint complaints were thought to be the most likely explanation. After childbirth, she presented to the outpatient clinic. The pain had spontaneously decreased. She still experienced pain, mainly at night, but sometimes also during the day. She still reported no pain in her arms or legs. Her family history was not contributory. She did not smoke or drink alcohol. She did not use any medication. Because she was breastfeeding her newborn baby, she did not take any pain medication.

The neurological examination showed no motor deficits. There was no clear deficit in sensory testing, but a slight decrease in vital sensibility in her arms and legs was suspected. Tendon reflexes were normal. Plantar reflexes were normal on both sides. The pain could not be provoked by manipulation of the spine. As the pain continued at night and no good explanation could be found, imaging of the spine was ordered. However, the a priori chance of finding a tumor was thought to be very low. An MRI scan of the thoracic spine showed an intramedullary, partly cystic/necrotic, partly solid tumor at levels C5–Th1. The solid part of the tumor was 3 cm long. It showed eccentric growth and was surrounded by edema. Using gadolinium, the solid part of the tumor showed enhancement, indicating a disruption of the blood-brain barrier (Fig. 1). The patient was referred to a neurosurgeon, who performed a debulking of the tumor. Histopathological examination showed tissue of high cellularity and pseudorosettes. There were hardly any mitoses seen, no necrosis, and some calcification. These findings are consistent with a WHO grade II ependymoma.

At follow-up, the patient had made a full recovery. Repeated MRI scans showed that the debulking was total, and there were no signs of recurrence.

### Discussion

We describe a patient who complained of isolated severe thoracic pain during pregnancy, with a spontaneous reduction in pain following delivery. The pain was caused by an intramedullary ependymoma. Lower back pain is very common during pregnancy; however, spinal pain – which can be provoked or increased in the supine position – should prompt the clinician to search for an alternative diagnosis as this can be the first sign of a spinal tumor.

Ependymoma is a primary tumor of the central nervous system and can occur anywhere along the ventricular system and spinal cord. Usually, ependymomas are intramedullary tumors. 46–75% of all ependymomas are localized in the spinal canal [1, 2].

Symptoms vary according to tumor localization. Common symptoms of spinal ependymomas include back pain, spasticity in the lower extremities, gait ataxia, sensory loss, and

paresthesias [1]. Our patient presented only with severe nightly pain during her pregnancy. The diagnosis of a spinal tumor during pregnancy is rare. It is estimated that 1 in 1,000–2,000 pregnancies is complicated by a tumor (regardless of the location or tumor type). The main tumor types that are found in the spine during pregnancy are giant cell tumors, hemangiomas, schwannomas, and metastases [4].

There are different theories as to why spinal cord tumors become symptomatic during pregnancy. Neuro-epithelial tumors (including ependymomas) have been proven to express the estrogen receptor ER- $\beta$  [4] and possibly progesterone receptors [5]. Estrogen and progesterone levels rise during pregnancy, and can accelerate the growth of the tumor. Also, increasing levels of growth factors and angiogenic factors during pregnancy can cause an acceleration of the tumor growth. Lastly, but probably most importantly, hemodynamic changes during pregnancy (i.e., increased blood flow volume through the vertebral venous plexus secondary to compression of the vena cava by the uterus) can cause an increase in spinal compression due to venous pooling [4]. This venous pooling might occur mainly at night, because the uterus exerts more pressure on the vena cava in the supine-horizontal position, possibly explaining why our patient experienced more symptoms at night.

However, approximately 45% of all pregnant women experience lower back pain during their pregnancy [6]. Thoracic or cervical pain is less frequent. Alternative diagnoses should be considered. Some of these are listed here:

#### *Vertebral Hemangioma*

Vertebral hemangiomas are common in the general population with a prevalence of 10–12%. Only 1% of these hemangiomas become symptomatic. Symptoms include back pain (54%) and neurological deficit (45%) when a spinal cord compression is present. Pregnancy is a known factor for a deterioration of vertebral hemangiomas, mainly because of venous hypertension due to the compression of the vena cava by the uterus. Approximately 89% of all vertebral hemangiomas that become symptomatic during pregnancy occur in the thoracic spine. Symptoms usually occur in the third trimester. Diagnosis is made by MRI of the spine and biopsy of the lesion. Neurological deficit often necessitates urgent intervention, namely cesarean section and laminectomy. Prognosis is usually good with full neurological recovery [7].

#### *Aortic Dissection*

Another cause of thoracic pain or pain between the shoulder blades, usually with an acute beginning, is aortic dissection. Aortic dissection during pregnancy is extremely rare, with 0.1–0.4% of all aortic dissections occurring during pregnancy. The risk of aortic dissection during pregnancy is estimated at 0.0004%. The main risk factor is connective tissue disease (Marfan's disease). Other risk factors include an aortic root diameter of  $\geq 40$  mm, hypertension, bicuspid aortic valve, and coarctation. Some believe that pregnancy alone is an independent risk factor. Diagnosis is made by computed tomography angiography, and treatment is usually the termination of the pregnancy by a cesarean section and surgical management of the dissection. Maternal and fetal/neonatal mortality are high, approximately 20 and 40%, respectively [8].

#### *Nontraumatic Spinal Epidural Hematoma*

Spinal epidural hematoma (SEH) is a well-known complication of epidural procedures, but, although it occurs rarely, it can also occur spontaneously. The pathophysiology of spontaneous SEH is unclear. Pregnancy might be a risk factor for spontaneous SEH, because of hemodynamic and possibly hormonal changes. Usually, there is an abrupt onset of neck or back

pain, followed by paresthesias, and sensory and/or motor deficits within hours to days. The thoracic part of the spinal cord is most commonly affected, giving rise to pain between the shoulder blades. Diagnosis is made by MRI. In case of severe neurological deficit, neurosurgical intervention should take place within 36–48 h after onset of symptoms to prevent definitive neurological sequelae, even though more than half of all patients will not fully recover [9]. On the other hand, when there is no or minimal neurological deficit, nonoperative management with close observation should be considered as an option, since spontaneous recovery has been described [10]. Neurosurgical intervention by means of laminectomy has been thought to be safe during pregnancy, so cesarean section prior to the surgery is not necessary in most cases [9].

## Conclusion

We report a patient with a rare presentation of ependymoma in the cervicothoracic spinal cord during pregnancy. Spontaneous improvement of pain following delivery does not always point towards a benign cause. Lower back pain is very common during pregnancy; however, spinal pain – which can be provoked or increased in the supine position – should prompt the clinician to search for an alternative diagnosis as this can be the first sign of a spinal tumor.

## Statement of Ethics

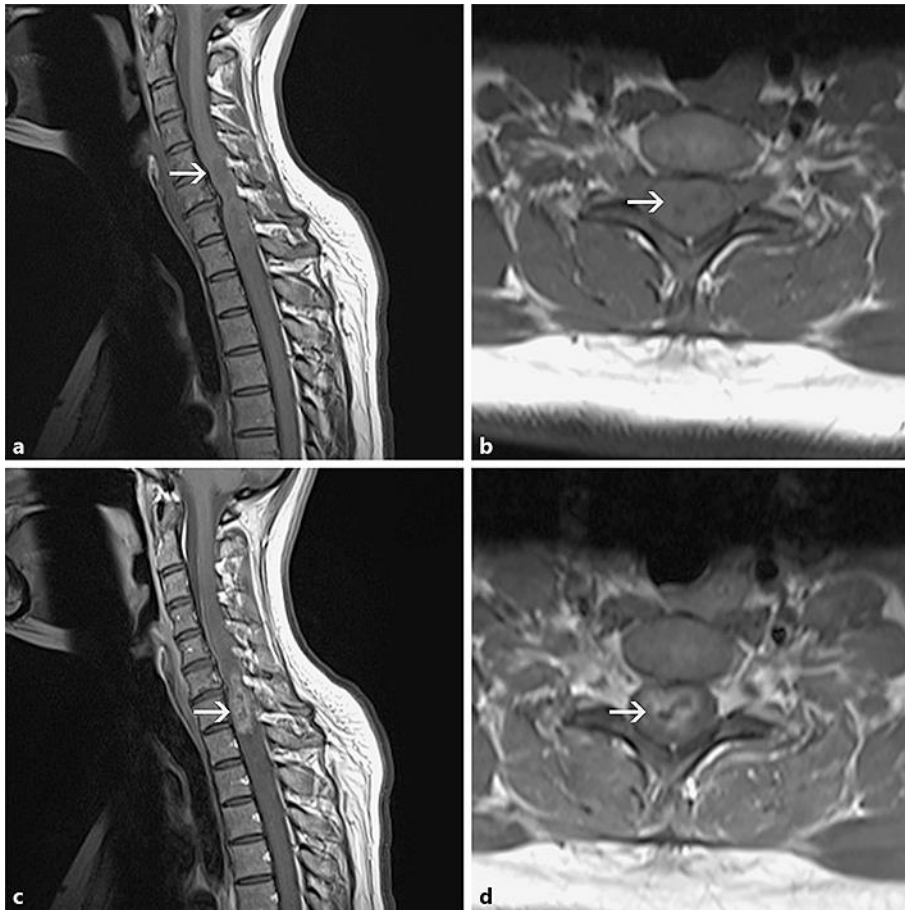
The patient consented to the publication of her case.

## Disclosure Statement

The authors declare that they have no conflicts of interest.

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**Fig. 1.** Sagittal T2 TIRM (a) and axial T1 (b) sequence shows a mixed solid and cystic intramedullary lesion (arrows) at C5–Th1 causing cord expansion that is heterogeneously hypointense on T1 and heterogeneously hyperintense on T2 TIRM with T2 hyperintensity extending cranially and caudally reflecting edema. Sagittal T1 postcontrast (c) and axial T1 postcontrast (d) shows a heterogeneous enhancing intramedullary mass (arrows) at C5–Th1.