Epidural anaesthesia for Caesarean section in a patient with severe Takayasu’s disease

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Takayasu’s arteritis or disease is a rare, idiopathic, chronic inflammatory disease which causes narrowing, occlusion or aneurysms of blood vessels. It preferentially affects large arteries such as the aorta and its branches and hence its alternative names of pulseless disease, occlusive thromboaoartopathy or aortic arch syndrome. Although most commonly found in oriental women, it occurs sporadically throughout the world. We present the case of an elderly primigravida with long-standing Takayasu’s disease complicated by hospital and needle phobia who underwent a successful Caesarean section under epidural anaesthesia. Her management is discussed in the light of current opinion regarding pregnancy and Takayasu’s disease.

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Case report

A 41-yr-old woman was referred to the antenatal clinic at 16 weeks’ gestation. The first symptoms of Takayasu’s disease had occurred at 15 yr of age but was not diagnosed as such until many years later. She was thought to have a connective tissue disorder and was treated empirically with steroids for several years. By the time of her pregnancy her disease had been considered quiescent for many years, but she was left with significant hypertension, extensive narrowing of her aorta and brachial vessels, and a pulseless left arm which, although ischaemic, was asymptomatic.

Before pregnancy her arterial pressure had been controlled at 160-190/0 mm Hg by a combination of antihypertensive agents, including lisinopril, amlodipine and doxazosin. Lisinopril was discontinued when pregnancy was confirmed because ACE inhibitors may be associated with congenital defects. Bisoprolol was added during the first trimester when systolic arterial pressure increased to 230 mm Hg. As her pregnancy progressed, control of arterial pressure within pre-pregnancy limits required greater doses of methyldopa in addition to a diuretic in the form of a frusemide-potassium combination.

The patient weighed 62 kg and was 1.65 m tall (BMI 22 kg m⁻²). She was clinically healthy with moderate exercise tolerance. She had no symptoms of decreased cerebral perfusion such as dizziness, fatigue or syncopal attacks. On physical examination there was no evidence of cardiac failure. Prominent but asymmetrical collapsing pulses were present in all limbs except her left arm. Bilateral carotid bruits and a diastolic murmur down the left sternal edge were heard on auscultation. Arterial pressure was 160/0 mm Hg in her right arm and 130/0 mm Hg in both legs, with no audible end-point to define diastolic pressure.

An echocardiogram confirmed moderate aortic regurgitation but good cardiac function with an ejection fraction of 79%. A colour Doppler duplex scan of her carotid arteries showed reduced blood flow with a grossly abnormal flow pattern suggestive of considerable intracranial resistance. A transcranial Doppler study of the middle cerebral arteries confirmed reduced blood flow, which was probably caused by intimal hyperplasia of the microcirculation. There was no evidence of stenosis or aneurysmal dilatation.

During the later stages of pregnancy, considerable efforts were made to reduce the patient’s anxiety regarding needles and hospitals, including the use of hypnotherapy, but with little success. The pregnancy was uneventful and progressed well without significant fetal or maternal problems until 32 weeks’ gestation. At this stage her weekly ultrasound scan showed a marked decrease in intrauterine growth, probably because of inadequate placental blood flow. A decision was made to deliver the infant as a semi-emergency procedure, only delaying by 24 h to allow administration of dexamethasone to accelerate fetal lung maturity.

Despite some reservations, and after much reassurance, the patient agreed that LSCS could be attempted under epidural anaesthesia. I.v. access was established after the use of topical local anaesthetic cream and an i.v. infusion of Hartmann’s solution was commenced. The patient was placed in the left lateral position and a 16-gauge Tuohy
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Discussion

Takayasu’s disease was first described in 1908 by two Japanese ophthalmologists, Takayasu and Onishi, who observed retinopathy occurring with absent limb pulses. Nowadays it is recognized as a rare (2–3 per million) inflammatory disease of the vascular tree, principally affecting major vessels such as the aorta, its branches and the pulmonary arteries. Although most commonly found in oriental women, it occurs sporadically throughout the world. It is more common in women than men (8:1), and the peak incidence is in the second and third decades, although a substantial minority, including this patient, present in their teens. The cause is unknown but it may have an autoimmune basis. Because it is more prevalent in women of childbearing age, sex hormones may be involved in the pathogenesis. A genetic predisposition has been seen in some families, the majority of which demonstrated BW52 human lymphocyte antigen (HLA).

The panarteritis of Takayasu’s disease results from intimal infiltration by lymphocytes and other inflammatory cells, with vessel wall elastic tissue being replaced by fibrous tissue. The arterial walls become progressively thickened and this leads to stenosis and occlusion, although dilatation and aneurysms may also occur. The disease is characterized classically by a triphasic course, commencing with an initial active phase of constitutional symptoms such as malaise, weight loss, fever, myalgias and arthralgias, and is associated with an increase in acute phase reactants (e.g. ESR). This progresses to a second stage with symptoms of cerebral, visceral or extremity ischaemia before resulting in a final ‘burnt-out’ quiescent phase of fibrosis. Unfortunately, these ‘typical’ patients are in the minority and the pattern, severity and progression of vessel involvement are extremely variable.

A substantial minority of patients with Takayasu’s disease are asymptomatic but approximately 60% suffer significant disability from complications, particularly hypertension and its consequences. Dizziness, vertigo and visual disturbances result from vertebral artery involvement; angina, cardiac and renal failure and intermittent claudication are also common. Treatment in the initial phase is with steroids (usually for 2–4 yr) with or without cytotoxic agents. This induces remission in approximately 80% of all patients although 50% of those subsequently relapse and require a further course of steroids. Subsequent treatment is limited to angioplastic or surgical correction of the stenoses or aneurysms. The disease tends to be self-limiting in 20% of patients while in a similar percentage the disease remains persistently active. There is also increasing evidence that occult inflammation may continue to occur despite clinical evidence of a remission. The overall 5-yr mortality is not clear but has been variously reported to be as low as 3% and as high as 35%. Survival has been estimated to be approximately 12–16 yr after diagnosis with the presence of retinopathy, aortic regurgitation and significant aneurysm formation being the worst prognostic indicators. The commonest causes of death are heart failure, myocardial infarction and stroke.

The severity of Takayasu’s disease can be classified in several ways, most commonly according to anatomical location and extent of disease, as determined by angiography and vascular signs and symptoms (such as the presence of bruits, symmetrical pulses, claudication, etc.). Ishikawa and Matsuura noted that in pregnancy, the degree of severity of retinopathy, secondary hypertension, aortic regurgitation and arterial aneurysm were particularly significant indicators of maternal outcome and he classified patients into four groups. Group I had none of the above complications, group Ia had one complication of mild severity, group Ib had one complication of marked severity and those with two or more complications were allocated to group III.

Although our patient had severe disease (group III), according to both established criteria and angiography (carried out 8 yr previously) and based on her hypertension and aortic regurgitation, she was largely asymptomatic (apart from headaches) throughout her pregnancy. This may
have been because of treatment with steroids when she had first presented more than 20 yr previously, which could have induced a remission and significantly limited her disease. Although the effect of pregnancy, labour and delivery is not known to alter disease activity, a substantial number of patients have worsening of complications, particularly cardiac decompensation. Fortunately, that did not occur in our patient.

Vaginal delivery, usually with epidural anaesthesia, is acceptable for patients in groups I and IIa, although the duration of the second stage is often deliberately shortened by instrumental delivery, particularly in hypertensive patients. Operative delivery is preferred for patients with stages IIb or III but is reserved for specific obstetric indications in less severely affected individuals. Its aim is to avoid the increase in blood volume and hence arterial pressure found during uterine contractions. In association with the increased cardiac output normally seen during pregnancy and labour, the likelihood of cardiac decompensation is increased further and is best avoided in these susceptible individuals.

Because of our patient’s hospital and needle phobia, a general anaesthetic was first thought to be the best option. This was previously the preferred method for operative delivery because of concern about hypotension caused by regional anaesthesia. However, both induction and extubation can involve substantial changes in arterial pressure, particularly with rapid sequence induction. More importantly, cerebral blood flow is difficult to assess under general anaesthesia. This was important in our patient, as her intracranial vessel resistance was so great that any decrease in arterial pressure could cause cerebral ischaemia. If a general anaesthetic is unavoidable in Takayasu’s disease, then EEG studies can be helpful, although transcranial Doppler may be preferable to monitor cerebral blood flow.

Although spinal anaesthesia for delivery of patients with Takayasu’s disease has not been reported (except for therapeutic termination of pregnancy in a patient with mild Takayasu’s disease) the use of epidural anaesthesia is becoming more accepted as hypotension is not commonly a problem and cerebral function can be assessed easily in an awake patient. Epidurals also allow parents to participate in the delivery of their baby and can provide good pain relief after operation, reducing the risk of further increases in arterial pressure. Miekle and Milne suggested caution in the use of epidural anaesthesia where there is a significant difference between arterial pressure in the upper and lower limbs. They contended that sympathetic block caused by the epidural may worsen already limited regional blood flow in an unpredictable ‘steal’ type manner. In our patient, there was a difference in systolic pressure of 30–40 mm Hg between the upper and lower limbs. However, in view of her high intracranial resistance, it was felt that the risk of exacerbating regional ischaemia was less than the advantages to be gained from an awake technique.

Patients with group III disease are more likely to have babies with intrauterine growth retardation and this was the indication for LSCS in our patient after the ultrasound scan at 32 weeks’ gestation. Intrauterine growth retardation is thought to be unrelated to the disease process behind Takayasu’s disease and more likely results from chronic systemic or renovascular hypertension limiting placental blood flow. Most of the pregnancies reported have continued to near term. This was not possible in our patient because of her age and the severity of her hypertension.

Placement of the epidural catheter and establishing adequate regional anaesthesia were uncomplicated. Arterial pressure was measured throughout non-invasively. Before surgery, a sphygmomanometer was used while during surgery and after operation an electronic method was used. In common with several case reports published previously, we saw a small decrease in arterial pressure after the first dose of local anaesthetic. This was corrected easily by a modest amount of ephedrine without the patient feeling dizzy, light-headed or nauseated. The patient’s arterial pressure remained unchanged during operation, even when Syntocinon was given. Ergometrine, although not needed in this case, is contraindicated because of its extreme effect on arterial pressure. Pulmonary oedema has been reported after delivery in Takayasu’s disease, although this is more likely in patients with poor cardiac function. For this reason, i.v. fluids were limited to just over 1 litre in our patient. Prophylactic antibiotics were given to prevent endocarditis and puerperal sepsis.

Invasive monitoring was considered and rejected, despite some reports suggesting that pulmonary artery flotation catheters, central venous pressure and direct measurement of arterial pressure by an arterial catheter may be of value in this situation. In the first instance, the patient was very reluctant to consent to such monitoring and fortunately, unlike many other patients with Takayasu’s disease, it was possible to measure arterial pressure consistently from the right brachial artery indirectly. Arterial pressure, as measured by sphygmomanometry, was the same as that measured by preoperative Doppler studies thus confirming its accuracy. This method was quick and effective and was, just as useful to examine the trend in arterial pressure changes. Moreover, there is increasing evidence from several case reports that arterial pressure generally remains unaffected in these patients undergoing surgery, regardless of the method by which arterial pressure is measured. This is perhaps not surprising given that the fixed rigidity of the vascular tree from fibrosis may be expected to prevent vasodilatation. As a precaution, however, more extensive monitoring was immediately available if required.

Our patient was monitored carefully during her stay in hospital, particularly in the first 24 h after delivery, as figures suggest that more than 60% of patients have postoperative problems, usually caused by inadequate control of arterial pressure, with heart failure or, devastating, frequently fatal, stroke in a small number of individuals. Fortunately, our patient had a smooth uncomplicated recovery throughout

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The overall maternal mortality rate of women with Takayasu’s disease is 4.8%26 and further pregnancies are not advised in patients with group IIb or III disease.

In summary, this patient is the oldest recorded primigravida to have a live birth with Takayasu’s disease. Her age and length and severity of Takayasu’s disease made her successful and uncomplicated Caesarean section under epidural anaesthesia particularly noteworthy. It also confirms the feasibility of epidural anaesthesia, even in the presence of needle and hospital phobia.

References

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