Surgery without Blood Transfusion for Giant Paraganglioma in a Jehovah’s Witness Patient

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Key Words
Giant paraganglioma · Jehovah’s witnesses · Blood transfusion refusal

Abstract
A retroperitoneal tumor was identified in a 57-year-old female belonging to Jehovah’s Witnesses during a health check. Subsequent examination led to the suspicion of a right pheochromocytoma. The patient wished to be treated by bloodless surgery and consulted our hospital after being refused surgery by several hospitals. She signed a liability waiver for blood transfusion refusal. After obtaining consent for diluted autotransfusion and preoperative administration of erythropoietin, the surgery was scheduled. The tumor was attached to the inferior vena cava and left renal vein and engulfed the right renal artery and vein. The tumor and right kidney were removed en bloc. Operative time was 8 h and 18 min, with 1,770 ml of blood loss. The histopathological diagnosis was paraganglioma with the normal adrenal gland within the border of the tumor. The patient was discharged from the hospital with no postoperative complications.

Introduction
At the present time (2013), 7.78 million Jehovah’s Witnesses are spread throughout 239 countries and regions and among them, blood transfusion refusal is common. We experi-
enced surgical treatment for giant paraganglioma without blood transfusion in a Jehovah’s Witness patient.

**Case Report**

In February 2011, a retroperitoneal tumor was suspected by a health check in a 57-year-old Japanese woman and subsequent examination led to the suspicion of a right pheochromocytoma. As the patient was a Jehovah’s Witness, she wished to undergo bloodless surgery and consulted our hospital in June 2011 after being refused surgery by several hospitals.

The characteristics of the patient recorded at the time of hospital admission were as follows: height: 154 cm, weight: 56 kg, blood pressure: 115/65 mm Hg and heart rate: 78 bpm and regular. The tumor was palpable in the right hypochondrium. There were no physical signs of Cushing’s syndrome such as moon facies or buffalo hump. There were no abnormal findings in the peripheral blood and biochemical tests. Endocrine tests revealed high levels of urinary normetanephrine (1.60 mg/day). There were no abnormal findings in any other items, including blood adrenaline, cardiac function, and pulmonary function.

Abdominal contrast-enhanced computed tomography (CT) revealed a retroperitoneal tumor (size: 12 cm) touching the upper pole of the right kidney with an uneven contrast-enhanced effect inside the tumor. The tumor had ill-defined borders with the surrounding liver, right kidney, inferior vena cava, right renal artery and vein, and left renal vein, which indicated adhesion (fig. 1). There was no clear lymph node metastasis or distal metastasis on imaging. The tumor had a low signal intensity on T1-weighted magnetic resonance images and high signal intensity on T2-weighted magnetic resonance images. Marked accumulation was found on $^{131}$I-metaiodobenzylguanidine scintigraphy.

The above findings were suggestive of a right adrenal pheochromocytoma. Abdominal contrast-enhanced CT findings revealed that tumorectomy required a right renal combined resection, sectioning of the right renal artery and vein, left renal vein, and inferior vena cava, and detachment of the tumor from the lower hepatic surface. Despite explaining the importance of blood to the patient once again as an outpatient, she did not give consent. However, we were able to gain her consent for the use of albumin preparations, intraoperative diluted autotransfusions, intraoperative recovery-type autotransfusions, the use of a heart-lung machine, and hemodialysis. A preoperation conference was held with the four hospital departments including anesthesiology, gastrointestinal surgery (because of the detachment of the tumor from the liver), and cardiovascular surgery (extracorporeal circulation is required when sectioning the inferior vena cava). The type of surgical procedure was confirmed, and the risks involved were sufficiently discussed. Then, the patient and her friends were provided the details of the surgery. The patient also acknowledged the risk of bleeding to death because of bloodless surgery, loss of renal function because of the removal of the right kidney and left renal vein, the possibility of hemodialysis, the risk of partial liver removal, and eventual exploratory laparotomy. After the patient understood all risks and consented, a written consent and waiver of liability for blood transfusion refusal were signed. In accordance with the surgical policy, an erythropoietin preparation, iron, and doxazosin mesylate were administered prior to surgery.

After the diagnosis of right pheochromocytoma, open surgery was conducted with an abdominal midline incision in August 2011. During operation, the tumor was found adhered to the peritoneum, diaphragm, lower hepatic surface, inferior vena cava, and right renal vein. It engulfed the right renal artery and vein.
First, the adhesion site of the tumor was detached from the lower hepatic surface. Partial hepatic resection was not necessary. Then, the anterior surface of the tumor was detached from the left renal vein, and the left renal vein was secured. Next, the right renal artery was tied, and blood flow to the right kidney was blocked. A clamp test was performed on the inferior vena cava and left renal vein, and as there were no changes in blood pressure, the bypass was considered adequate. Next, the tumor was very carefully detached from the inferior vena cava, which was tied to protect the right renal vein as tumor detachment progressed. Lastly, the tumor vessels flowing into the inferior vena cava were tied, and both the right kidney and tumor were removed. The inferior vena cava and left renal vein were preserved. The operative times were 8 h and 18 min and blood loss was 1,770 ml.

The resected specimen (size: 10.5 × 11.9 cm) was macroscopically brown in appearance with multiple cysts. There was no continuity of the tumor in the normal adrenal gland, and it was diagnosed as paraganglioma (fig. 2).

On histological findings, the tumor consisted of a hyperplasia of cells with basophilic cytoplasm and was arranged in an alveolar pattern around fibrous blood vessels (fig. 3). Histologically, there was no clear continuity of the tumor in the adrenal gland tissue, and an extra-adrenal paraganglioma was diagnosed. No invasion into the right adrenal gland or right kidney was observed.

Following surgery, the patient progressed without any major changes in blood pressure or other complications and the high urinary normetanephrine level returned to normal. After discharge, the patient continues to have follow-ups at her home hospital.

Discussion

In 1869, a small Bible study group was founded in the state of Pennsylvania in the US. The group preached that the Bible should be strictly followed and took a different position from the traditional Catholic doctrine. Through the publication of journals and active missionary work, the group gradually gained widespread support throughout the US and took on the characteristics of a Christian denomination. In 1931, they began calling themselves the Jehovah’s Witnesses (The Watchtower, A New Name, October 1, 1931, pp 291).

In 1948, Jehovah’s Witnesses recommended blood transfusion refusal on the basis of citations from teachings in the Old Testament such as ‘you must not eat the blood of any living creature’ (Awake!, October 22, 1948, pp 12). They preach that eating blood is connected to blood transfusions because ‘receiving glucose solution transvenously is called transvenous nutrition.’ This refers to the feeding of patients through an intravenous drip. Therefore, blood transfusion is the same as eating blood from the vein (The Watchtower, July 1, 1951, pp 415). Thereafter, Jehovah’s Witnesses refused blood transfusions in medical care and declined surgery or treatment when a large volume of blood was expected to be lost.

In the US at the start of the 1960s, medical, legal, and ethical issues involving cases of blood transfusion refusal sparked many debates and absolute bloodless surgery began to be performed on the basis of the rights of the patient [1, 2]. Most noteworthy are the results of Ott et al. [3] who performed bloodless cardiovascular surgery on 542 Jehovah’s Witness patients between 1957 and 1977. They reported that only 3 of 542 patients died during surgery because of blood transfusion refusal. Since then, many reports have described surgical cases of Jehovah’s Witness patients in which more than 1,000 ml of blood loss is expected, such as open heart surgery in the field of cardiovascular surgery [4, 5], hepatic
resection in the field of gastrointestinal surgery [6] and total hip replacement in the field of orthopedics [7].

To our knowledge, there has only been one reported case that describes surgery of a Jehovah’s Witness patient with a large pheochromocytoma in Japan [8]. Ito et al. [8] reported on a 59-year-old woman belonging to the Jehovah’s Witnesses who was diagnosed with pheochromocytoma in the left adrenal gland, measuring 11 cm in diameter. They preoperatively administered erythropoiesis-stimulating agents and alpha-1-blocker to manage anemia and maintain total blood volume and underwent left adrenalectomy without blood transfusion (the operative time was 4 h and 42 min and total volume of blood lost was 335 ml).

While addressing the issue of blood transfusion refusal has become a problem, the Japanese joint committee on blood transfusion refusal because of religious beliefs published the ‘Guidelines on Blood Transfusion on Religious Grounds’ in 2008 [9]. According to these guidelines, patients aged ≥18 years with the ability to make judgments and who refuse blood transfusions should be made aware of the importance of blood transfusions, risks involved in refusing them, and available autotransfusion options. If they still refuse, then blood transfusion cannot be performed. In the event that an absolute bloodless surgery is deemed possible, the surgery may proceed after the patient signs the ‘Certificate of Refusal Blood Transfusion and Exemption from Liability’, but if bloodless surgery is deemed difficult, then the patient may be asked to change doctors or be shown possible options to deal with blood transfusion refusal [9].

When performing absolute bloodless surgery, the suitability must be carefully examined, and high-risk patients should be examined by an ethics committee. The cooperation of other departments should be requested as needed, and medical professionals should be prepared for any potential issues that may occur during surgery.

About 7.78 million Jehovah’s Witnesses are spread throughout 239 countries and regions and in Japan, there are approximately 210,000 adherents (Watch Tower Bible and Tract Society of Pennsylvania, Yearbook of Jehovah’s Witnesses, 2013). Blood transfusion refusal because of religious beliefs is a problem that any doctor may face and requires the development and dissemination of legislation.

In conclusion, we treated a case of retroperitoneal paraganglioma (size: 12 cm) by conservative blood surgery because of the religious beliefs of the patient. Patients who refuse blood transfusions because of religious beliefs should be treated in accordance with the ‘the Refusal of Blood Transfusion on Religious Grounds’ in Japan. However, the decision of performing bloodless surgery is left up to the discretion of each individual physician because there are no clear criteria for the procedure. Bloodless surgery entails medical and legal risks and places a great burden on medical practices. Blood transfusion refusal because of religious beliefs requires further development and dissemination of legislation.

Disclosure Statement

The authors have no conflicts of interest to disclose.
References


Fig. 1. CT scan of the abdomen showing a right retroperitoneal mass. a Axial section and b coronal section.
Fig. 2. Macroscopic finding of a surgical specimen.

Fig. 3. Microscopic finding of the tumor. A polygonal basophilic atypical cell proliferated in a rich vascular network (HE stain, original magnification ×100).