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5 **Cause of postprandial vomiting - A giant retroperitoneal ganglioneuroma**
6 **enclosing large blood vessels: A case report**

7

8 Zheng X *et al.* A giant retroperitoneal GN

9

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11

12 **Abstract**13 **BACKGROUND**

14 Ganglioneuroma (GN) is a rare neurogenic tumor that accounts for about 0.1%-
15 0.5% of all tumors of the nervous system. It originates from neural crest cells.
16 GN has no specific clinical symptoms or laboratory findings, which leaves it
17 easily overlooked and misdiagnosed as other tumors. Retroperitoneal GN with
18 very large volume and vascular penetration is extremely rare.

19

20 **CASE SUMMARY**

21 We present the imaging and pathological findings of a giant retroperitoneal GN
22 in a child. A 4-year-old boy had suffered from postprandial vomiting for more
23 than 6 mo with no precipitating factors. Abdominal computerized tomographic
24 examination showed a giant cystic mass in the retroperitoneal area. After
25 injection of contrast agent, the mass showed heterogeneous enhancement.
26 Surgery with local excision of the mass was performed to address the
27 embedded abdominal blood vessels, and the histopathological and
28 immunohistochemical diagnosis of the mass was GN. Postprandial vomiting
29 was relieved, and no complications occurred after the operation.

30

31 **CONCLUSION**

32 In the diagnosis of giant retroperitoneal hypodense masses in children, GN
33 should be considered if the mass presents delayed enhancement, punctate
34 calcification, and vascular embedding but no invasion. Pathology is the
35 golden standard for the diagnosis of GN, and surgical excision is the optimal
36 treatment for GN.

37

38 **INTRODUCTION**

39 Ganglioneuroma (GN) is a benign neurogenic tumor that accounts for more
40 than 0.1%-0.5% of all tumors of the nervous system^[1]. It commonly occurs in
41 the mediastinum (41.5%) and retroperitoneum (37.5%)^[1-3]. Patients are usually
42 asymptomatic, and GN is often found via physical examination. However, the
43 tumor can press against the adjacent organs and cause complications if it
44 reaches a great enough size. Here, we introduce a case of a 4-year-old boy with
45 giant retroperitoneal GN associated with postprandial vomiting enclosing the
46 main abdominal vessels. To our knowledge, existing reports mention only
47 small arteries passing through GN, and there is no previous work reporting
48 GN enclosing so many large arteries. The aim of our report is to present a rare
49 case and discuss the related clinical, imaging, and pathological features.

50

51 **CASE PRESENTATION**

52 *Chief complaints*

53 A 4-year-old boy was admitted to our hospital with a history of postprandial
54 vomiting for more than 6 mo without precipitating factors and abdominal pain,
55 chill, fever, or anal cessation of exsufflation or defecation.

56

57 *History of present illness*

58 He had poor appetite but no weight loss during his illness.

59

60 ***History of past illness***

61 The child was of full-term natural delivery with normal feeding and timely
62 vaccination.

63

64 ***Personal and family history***

65 He had no previous or family history of similar illnesses.

66

67 ***Physical examination upon admission***

68 Physical examination showed abdominal bulge and intestinal type in the upper
69 abdomen. The epigastrium showed a palpable mass with poor mobility; the
70 lump was hard but was associated with no tenderness.

71

72 ***Laboratory examinations***

73 The laboratory examinations (including blood and urine routine tests,
74 coagulation function tests, liver and kidney function tests, and tumor markers)
75 were normal.

76

77 ***Imaging examinations***

78 The pre-contrast computerized tomographic (CT) scan presented a
79 retroperitoneal giant hypodense mass with a size of 10.7 cm × 17.3 cm × 15.5
80 cm and a CT value of 33.8 HU. Spotted calcification with irregular distribution
81 was noted in the mass. The tumor showed inhomogeneous flocculent
82 enhancement in the venous phase and further enhancement in the delayed
83 phase after intravenous injection of contrast agent (Figure 1). We observed that
84 the stomach had been pushed forward and displaced, which was also
85 considered the main cause of postprandial vomiting for the patient. The tumor
86 surrounded the ³ celiac trunk, hepatic artery, splenic artery, superior mesenteric

87 artery, bilateral renal artery, and portal vein. Nevertheless, the enclosed
88 vascular lumen did not become narrowed or distorted (Figure 2 and 3). Of
89 course, this created a difficult problem for the patient's surgical treatment, such
90 that the surgeon was only able to remove the relatively non-vascular part of the
91 tumor.

92 Histopathology revealed the spindle cell tumor with nerve fibers and
93 ganglion cells. Immunohistochemical investigation showed the tumor cells
94 expressed NSE (diffuse +), Syn (+), S100 (diffuse +), and NF (diffuse +) and
95 were negative for GFAP, CR, P53, and SMA. The Ki-67 proliferation index was
96 5%.

97

98 FINAL DIAGNOSIS

99 Retroperitoneal GN.

100

101 TREATMENT

102 The patient underwent partial resection of two retroperitoneal tumors about
103 6.0 cm × 3.0 cm × 2.0 cm and 6.0 cm × 5.0 cm × 4.0 cm in size.

104

105 OUTCOME AND FOLLOW-UP

106 Postprandial vomiting was relieved, and no complications occurred. The
107 patient was discharged 10 d after surgery. The patient's parents declined
108 follow-up after discharge and did not disclose their reasons.

109

110 DISCUSSION

111 Retroperitoneal GNs account for 37.5% of all GNs and about 0.72%-1.6% of
112 primary retroperitoneal tumors^[1-3]. GNs can also occur in the vertebra, neck,
113 and cerebellopontine angle region (trigeminal); all three of which are relatively
114 rare^[4-8]. GNs are found incidentally in most cases and manifest as

115 asymptomatic masses^[2,9,10]. The tumor could cause some complications if it
116 becomes large enough to press against the adjacent organs. In our case, the
117 stomach dilated after meals, peristalsis became limited after meals due to the
118 compression from the GN, and food could not easily enter the duodenum,
119 causing postprandial vomiting. Occasionally, GNs occurring in the adrenal
120 gland can secrete vasoactive intestinal peptides, dopamine, and cortisol, which
121 lead to diarrhea, hypertensive crisis, and male-like metabolic disorders in
122 women^[11-13].

123 GNs are mainly composed of ganglion cells, mucus matrix, nerve fibers, and
124 mature Schwann cells, and the first two of them are characteristic components
125 in histopathology^[14,15]. The pathological features of GN are closely related to its
126 CT findings. The presence of mucus matrix in tumors determines the
127 hypodensity on plain CT scans. The mucus matrix has been found to delay the
128 absorption of contrast agents, which leads to the delayed enhancement of
129 GNs^[16,17]. This case was misdiagnosed as retroperitoneal cystic lymphangioma
130 initially. Similar to this case, cystic lymphangioma tends to occur in children
131 and presents as a large, well-defined low-density mass in the
132 retroperitoneum^[18], which can also wrap around blood vessels without
133 distorting them, while a retroperitoneal GN of such a large size as this case is
134 rare and it is predominant in adults. However, in this case, flocculent and strip
135 delayed enhancement was observed, which was consistent with the
136 enhancement of GN, while cystic lymphangioma was generally not enhanced,
137 which was the most important distinguishing point between them.
138 Calcifications have been noted in 20%-60% of GNs, and most of them are
139 punctate, which is also one of the differences between GN and
140 neuroblastoma^[19,20]. Punctate calcification with scattered distribution was also
141 noted on plain CT images in this case. Ko *et al*^[14] and Duffy *et al*^[21] suggested
142 that the presence of fat components in GN may be one of the characteristics of

143 GN, but their sample size was too small to verify this, and we did not notice
144 any fat replacement in our case. Some scholars have proposed that the blood
145 vessels are often surrounded or compressed by GNs instead of being invaded,
146 although most of them are small vessels^[16,22], and this finding further suggested
147 that GNs are benign. In this case, GN enclosed all major abdominal vessels,
148 including the celiac trunk, hepatic artery, splenic artery, bilateral renal artery,
149 superior mesenteric artery, inferior vena cava, and portal vein. None of the
150 enclosed vessels had a narrowed lumen or filling defect, which provided more
151 reliable support for these scholars' views. However, there are some reports
152 suggesting that GNs could behave aggressively, and recurrence or malignant
153 transformation^[2,23,24] and complete surgical excision are the most optimal
154 choice for the treatment^[8,25]. Therefore, patients with GN still need long-term
155 radiological follow-up. In addition, some scholars have put forward a different
156 view, arguing that incomplete resection of GN ⁴ does not increase the risk of
157 progression if residual tumors are less than 2 cm in diameter^[10]. We believe that
158 it is important to determine whether the tumor is invasive before surgery and
159 assess pathology to select the most suitable surgical method, an area in which
160 we believe the current research into GN is lacking.

161 In conclusion, GNs appear as hypodense masses on plain scans and
162 present delayed and mild enhancement on contrast enhancement. Pathology is
163 the gold standard for the diagnosis of GN, and mucous matrix and ganglion
164 cells are their important features. Surgical excision is the best treatment for GN,
165 and postoperative radiotherapy and chemotherapy are unnecessary. However,
166 in cases where it is difficult to completely dissect the tumors and blood vessels,
167 partial resection could still relieve the pressure symptoms created by the
168 tumors. Long-term radiological follow-up after the operation is necessary, even
169 though the biological behavior of GN is benign, because there is still a tendency
170 to be malignant, especially in patients with GN who have undergone only local

171 resection.

172

173 CONCLUSION

174 Retroperitoneal GN, which is huge and encloses a large number of blood
175 vessels without invading, is rare in clinical settings. Although some previous
176 studies maintain that complete resection of the tumor is not necessary, finding
177 suitable ways of assessing the invasiveness of GN after pathology and
178 radiology still needs further discussion and study.

179

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183

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271

272 **Figure 1 Multiphase enhanced computerized tomographic images of the**
 273 **patient.** A: Giant retroperitoneal hypodense mass in pre-contrast and punctate
 274 calcification could be noted; B: The hypodense mass without enhancement in
 275 arterial and venous phase, and the stomach and kidneys were compressed and
 276 displaced; C: The mass presented mild enhancement in venous phase; D: The
 277 mass showed patchy and flocculent enhancement in delay phase, and the
 278 extent of enhancement was greater than that of the venous phase.

279

280 **Figure 2 The computerized tomographic images of tumors surrounding**
281 **blood vessels.** A-D: Tumor-enclosed arteries: hepatic artery (A), splenic artery
282 (A, B), bilateral renal artery (C), superior mesenteric artery (D); E, F: Tumor-
283 enclosed veins: Portal vein (E) and inferior vena cava (F).
284

285 **Figure 3 Coronal and sagittal of the computerized tomographic images. A, B:**
286 Tumor-enclosed arteries; C: Tumor-enclosed veins.
287

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