CASE REPORT

RJME Romanian Journal of Morphology & Embryology http://www.rjme.ro/

Spontaneous rupture of immature gastric teratoma with hemoperitoneum in a newborn with 3-year follow-up

CHUL KYU ROH[1)], MIN JUNG JUNG[2)], JIYOON KIM[2)], SUSIE CHIN[2)], AHRIM MOON[2)]

[1)]*Department of Surgery, Ajou University School of Medicine, Suwon, Korea*

[2)]*Department of Pathology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon-si, Gyeonggi-do, Korea*

Abstract

Among the subtypes of germ cell tumors, teratomas are the most frequent in the pediatric population and commonly occur in the sacrococcygeal region and the gonads. Less than 1% of all teratoma are found in abdominal organs including the stomach, liver, and kidney. Gastric teratomas are very rare tumors predominantly found in infants. Moreover, an immature gastric teratoma is exceptionally rare. Here, we present a case of immature gastric teratoma with spontaneous rupture in a newborn who was preoperatively diagnosed with neuroblastoma. On the first day after birth, the neonate presented with progressive abdominal distension accompanying respiratory distress. A firm mass was detected during a physical examination of the abdomen. An emergency exploratory laparotomy revealed hemoperitoneum resulting from a rupture of the tumor located in the posterior wall of the gastric antrum. Complete resection of the tumor and gastroduodenostomy were performed. The pathology evaluation revealed a grade 3 immature gastric teratoma with no malignant components. The patient was treated with adjuvant chemotherapy to prevent recurrence, since the tumor was ruptured in the abdominal cavity and the level of alpha-fetoprotein was decreased but still remained high above the normal range after surgery. In conclusion, physicians should be aware of the existence of gastric teratoma as the differential diagnosis of a huge abdominal mass in infants, especially neonates. Complete surgical removal of the tumor and long-term follow-up has been adopted as the standard management for immature gastric teratoma, although there has been controversy with adjuvant chemotherapy.

Keywords: stomach neoplasm, teratoma, newborn, rupture, hemoperitoneum.

Introduction

Germ cell tumors are composed of tissues from more than one of the embryonic germ layers and usually contain foreign tissues to the organ or anatomic site of origin. Among germ cell tumors, teratomas are the most common subtype in the pediatric population. The most commonly involved sites of teratomas in the pediatric population are the sacrococcygeal region and the gonads [1]. Primary gastric teratomas are extremely rare neoplasms (less than 1% of all teratomas in children) and the vast majority of reported cases are histologically mature teratomas [2, 3]. Other childhood teratomas occur mainly in female, while gastric teratomas are markedly predominant in boys [4]. Eustermann & Sentry first reported gastric teratoma in 1922 [5]. After that report, few cases have been reported as an immature gastric teratoma [3].

Aim

We report the first case of ruptured immature gastric teratoma accompanied by hemoperitoneum in a neonate.

Case presentation

This case report was approved by the Institutional Review Board of Soonchunhyang University Bucheon Hospital, Korea (IRB No. 2019-04-010). Informed consent was waived by the Institutional Review Board through full anonymization of patient information. A newborn boy was born at 41+3 weeks' gestation by Caesarean section. His birth weight was 3600 g. On the first day after birth, the neonate showed progressive abdominal distension with respiratory distress. Physical examination revealed abdominal fullness and a firm palpable mass along the left upper and lower quadrants. Plain radiography showed huge soft tissue mass with inner calcification in the left abdomen and displaced bowel loop to the right (Figure 1A). Ultrasonography (US) demonstrated a heterogeneous huge solid lesion with cystic change and irregular calcifications in the abdominal cavity (Figure 1B). The ultrasonographic diagnosis was congenital neuroblastoma arising from the left adrenal gland and the differential diagnosis was teratoma with suspicious partial rupture. The patient underwent exploratory laparotomy through a right supra-umbilical transverse incision on the first day after birth in Soonchunhyang University Bucheon Hospital. On the opening of the abdomen, hemoperitoneum was found. The huge tumor was arising from the posterior wall of gastric antrum combined with focal rupture. There was no perforation of the stomach. After complete resection of the tumor, intestinal continuity was restored by gastro-duodenostomy. On a postoperative day 9, oral feedings was started. The patient was discharged on a postoperative day 18 on full strength formula without complications.

In the pathology assessment, the huge exophytic gastric tumor was measured as 10×8×7 cm in size and weighted 288 g. The cut surface of the tumor revealed solid and cystic appearance with rupture (Figure 2). A total of 12 slides were prepared for the tumor, and the variable amounts of neural tissue were scattered in 10 of 12 slides.

Ectodermal derivatives, such as skin and cutaneous appendages as well as brain tissue, mesodermal derivatives, such as cartilage and adipose tissue, and small amounts of endodermal derivatives, such as respiratory epithelium, were observed in varying proportions, and the degree of maturation also varied. In all 10 slides with brain tissue, immature neuroepithelial tubules and rosettes were observed, even up to four low-power fields in one slide. Cellular neuroglial tissue without distinct neuroepithelial tubules was frequently observed. These immature neuroectodermal tissues accounted for about 20% of the tumor. Immunohistochemical staining for glial fibrillary acid protein (GFAP) showed positive findings in mature nerve tissue, and GFAP expression in immature neuroectodermal component was weak and focally positive. There were no malignant components including yolk sac tumor. The tumor was completely resected with negative resection margins. Based on the results of the imaging studies and surgery along with focal rupture of the tumor, the clinical stage was stage II. The final pathological diagnosis was immature gastric teratoma, grade 3 according to the Norris grading system (Figure 3) [6]. Neonatal serum alpha-fetoprotein (AFP) level was first evaluated on day 5 after birth and was elevated to 17 111 ng/mL. AFP level was decreased to 149.5 ng/mL on six weeks after birth. Human chorionic gonadotrophin was within normal range on day 5 after birth (3.4 mIU/mL) and decreased to less than 0.1 mIU/mL on six weeks after birth. The patient was treated with adjuvant four cycles of Bleomycin, Etoposide and Cisplatin chemotherapy, since AFP remained a markedly high level above the normal range and grade 3 immature teratoma was ruptured in the abdominal cavity. Serial abdominal US, magnetic resonance imaging, and serum AFP assessment were performed for evaluation of recurrence. The patient has no tumor recurrence for three years after surgery so far.

Discussions

The noteworthy reasons for reporting our case are the following: (*i*) a grade 3 immature gastric teratoma in a newborn is extremely rare; (*ii*) to our knowledge, this is the first report of ruptured gastric teratoma associated with hemoperitoneum; (*iii*) after complete surgical resection, adjuvant chemotherapy was performed to prevent recurrence in grade 3 immature gastric teratoma without malignant components.

Gastric teratomas most often occur in infants and children, though it can occur at any age. Although the exact etiology of gastric teratomas is not known, germ cell theory is generally accepted, which hypothesizes that extragonadal teratomas originate from migration of totipotent germ cells. Gastric teratomas differ from other teratomas in that it has a male preponderance, has an excellent prognosis, and is not associated with other congenital malformations. Gastric teratomas, especially those that are mostly matured, usually exhibit an exogastric growth pattern and typically present as palpable abdominal mass or abdominal distension during the first year of life [7–9]. An immature gastric teratoma in a newborn is extremely rare and clinically present as abdominal distension with respiratory distress. An immature gastric teratoma in a newborn may be a part of the spectrum of congenital tumors [10–14]. Since the presence of teeth or bone, which is pathognomonic for teratoma, is less frequently present in gastric teratoma, most gastric teratomas are frequently diagnosed preoperatively as a neuroblastoma, Wilms' tumor, mesoblastic nephroma, rhabdomyosarcoma, liposarcoma, or retroperitoneal teratoma [8, 15–17].

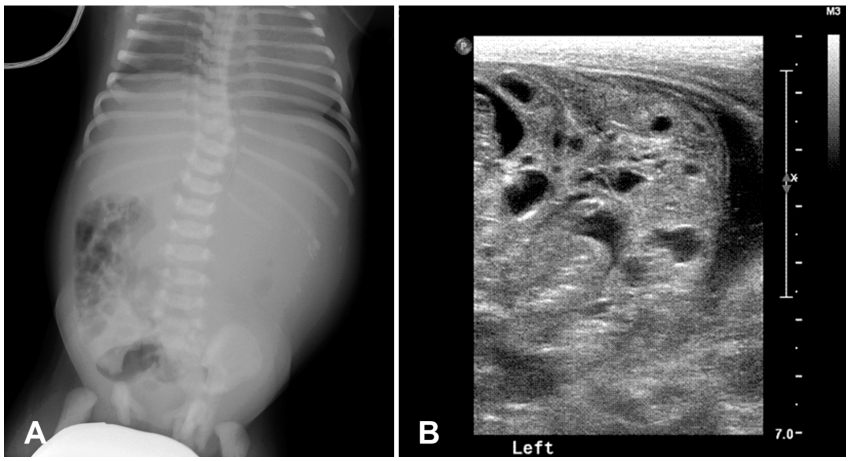

Figure 1 – ***Preoperative radiological evaluations: (A) Plain radiography revealed a huge soft tissue mass with inner calcification in the left abdomen and displaced bowel loop to the right; (B) Abdominal ultrasonography demonstrated a huge solid heterogeneous lesion with cystic changes and irregular calcifications in the abdominal cavity.***

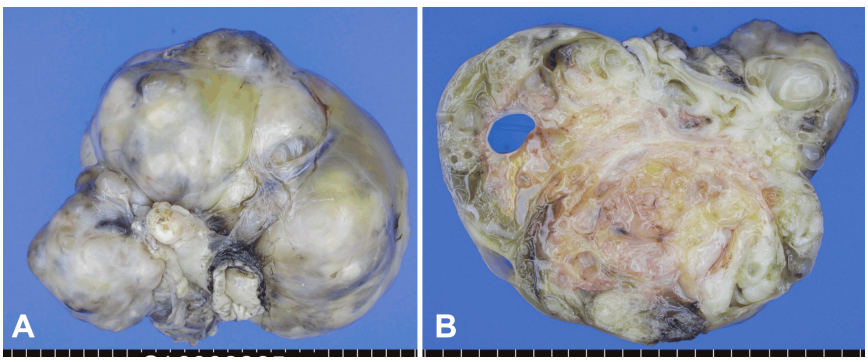

Figure 2 – ***Photographs of the immature gastric teratoma: (A) Gross specimen; (B) Cross section of the gross specimen.***

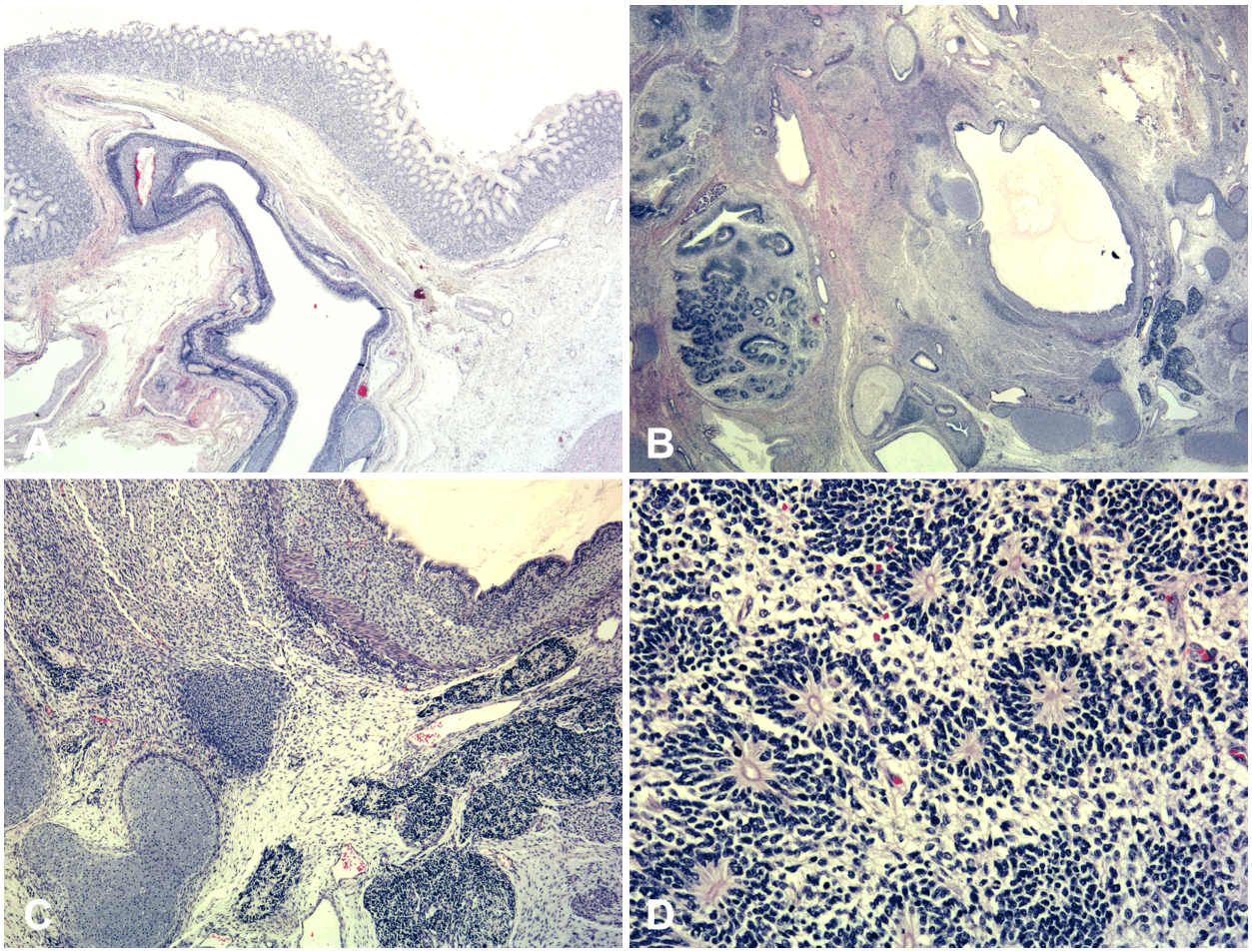

Figure 3 – ***Microscopic evaluations of the immature gastric teratoma: (A) Scan power view of the immature gastric teratoma showing normal gastric epithelium with ectodermal elements; (B and C) Low-power view showing ciliated columnar epithelium, immature cartilage, and glial tissue; (D) High-power view showing neuroectodermal rosettes. Hematoxylin–Eosin (HE) staining: (A) 20×; (B) 10×; (C) 40×; (D) 200×.***

In our case, we excluded mesoblastic nephroma since the kidneys appeared normal *via* US. Neuroblastoma with extragastric extension is rare. Obviously, there were abundant embryonic-type neural tissues in our case. However, they were admixed with other typical teratomatous elements, with varying degrees of maturation. These histopathological (HP) findings and low expression of GFAP were consistent with the diagnosis of immature gastric teratoma. GFAP expression in nerve tissue of teratoma is a meaningful indicator for differentiating mature and immature teratomas. Practically, the HP findings described above, with results of a GFAP immunostaining, can help to classify the teratoma and to differentiate neuroblastomas, sarcomas, as well as primitive or embryonal neoplasms arising within an immature teratoma.

Immature gastric teratoma containing neuroepithelial components may be potentially malignant [18]. The malignant component of tumors with mixed histology that contain teratomas is usually a yolk sac tumor. The presence of malignant yolk sac components is related with elevated levels of serum AFP, which is the only alerting sign [19, 20]. In our case, although markedly elevated AFP level was identified, we cannot detect the microscopic foci of yolk sac tumor despite meticulous pathological examination. In the gastric teratoma, an increase in AFP level without yolk sac tumor is associated with normal synthesis of AFP in gastrointestinal tract besides the yolk sac and liver during ontogenesis [21]. Although AFP is the most useful for the diagnosis and follow-up of germ cell tumors, it may not be a predictive marker of the malignant potential of immature gastric teratomas [22, 23].

Complete surgical resection with tumor-free margins remains the gold standard treatment in the management of gastric teratoma [24]. Gastric teratomas may be a pedunculated requiring limited or partial gastrectomy or may be extensively broad-based requiring rarely total gastrectomy [7]. Although adjuvant chemotherapies are not recommended for patients with completely resected grades 2 and 3 immature teratoma, there is still no established consensus on the optimal treatment in these groups [7, 25].

Conclusions

Immature gastric teratomas are rare tumors especially seen in neonates. An accurate and differential diagnosis from other more common causes of intra-abdominal neoplasm in neonates is required. Since serum AFP level may not predict malignant potential, the specimen should be thoroughly sampled to avoid missing additional malignant components. Complete surgical resection is the treatment of choice and a long-term follow-up with serial evaluation of serum AFP level is required to detect recurrences that

can present even after a decade of apparently complete tumor resection.

Conflict of interests

The authors have no potential conflict of interests to disclose.

Acknowledgment of research support

This work was supported by the Soonchunhyang University Research Fund.

Informed consent

Approval of Ethical Committee from Soonchunhyang University Bucheon Hospital, Korea was requested and obtained for using data from patient's file and publication as scientific article.

References

[1] Göbel U, Calaminus G, Engert J, Kaatsch P, Gadner H, Bökkerink JP, Hass RJ, Waag K, Blohm ME, Dippert S, Teske C, Harms D. Teratomas in infancy and childhood. Med Pediatr Oncol, 1998, 31(1):8–15. https://doi.org/10.1002/(sici)1096-911x(199807)31:1<8::aid-mpo2>3.0.co;2-h PMID: 9607423

[2] Grosfeld JL, Ballantine TV, Lowe D, Baehner RL. Benign and malignant teratomas in children: analysis of 85 patients. Surgery, 1976, 80(3):297–305. PMID: 960000

[3] Parvin S, Sengupta M, Mishra PK, Chatterjee U, Banerjee S, Chaudhuri MK. Gastric teratoma: a series of 7 cases. J Pediatr Surg, 2016, 51(7):1072–1077. https://doi.org/10.1016/j.jpedsurg.2016.01.002 PMID: 26850909

[4] Azpiroz J, Valle E, Herberth A, Carrillo V, Vaca R. Gastric teratoma in infants. Am J Surg, 1974, 128(3):429–432. https://doi.org/10.1016/0002-9610(74)90186-x PMID: 4547215

[5] Eusterman GB, Sentry EG. Benign tumors of the stomach: report of twenty-seven cases. Surg Gynecol Obstet, 1922, 34:372–378.

[6] Norris HJ, Zirkin HJ, Benson WL. Immature (malignant) teratoma of the ovary: a clinical and pathologic study of 58 cases. Cancer, 1976, 37(5):2359–2372. https://doi.org/10.1002/1097-0142(197605)37:5<2359::aid-cncr2820370528>3.0.co;2-q PMID: 1260722

[7] Saleem M, Mirza B, Talat N, Sharif M. Gastric teratoma: our 17 year experience. J Pediatr Surg, 2018, 53(2):234–236. https://doi.org/10.1016/j.jpedsurg.2017.11.010 PMID: 29229478

[8] Gore MD, Fernbach SK. Case 52: gastric teratoma. Radiology, 2002, 225(2):497–499. https://doi.org/10.1148/radiol.2252010498 PMID: 12409586

[9] Hasan R, Monappa V, Kumar S, Kumar V. Large gastric teratoma: a rare intra-abdominal mass of infancy. Oman Med J, 2016, 31(3):231–234. https://doi.org/10.5001/omj.2016.44 PMID: 27162596 PMCID: PMC4852076

[10] Akram M, Ravikumar N, Azam M, Corbally M, Morrison JJ. Prenatal findings and neonatal immature gastric teratoma. BMJ Case Rep, 2009, 2009:bcr10.2008.1050. https://doi.org/10.1136/bcr.10.2008.1050 PMID: 21686482 PMCID: PMC3028133

[11] Herman TE, Siegel MJ. Congenital gastric teratoma. J Perinatol, 2008, 28(11):786–787. https://doi.org/10.1038/jp.2008.85 PMID: 18974753

[12] Jeong HC, Cha SJ, Kim GJ. Rapidly grown congenital fetal immature gastric teratoma causing severe neonatal respiratory distress. J Obstet Gynaecol Res, 2012, 38(2):449–451. https://doi.org/10.1111/j.1447-0756.2011.01728.x PMID: 22229956

[13] Aihole JS, Babu MN, Jadhav V, Javaregowda D. Gastric teratoma: an unusual presentation and location. Indian J Med Paediatr Oncol, 2017, 38(4):563–565. https://doi.org/10.4103/ijmpo.ijmpo_182_16 PMID: 29333033 PMCID: PMC5759085

[14] Kumar S, Yadav H, Rattan KN, Srivastava D, Chandana A, Prakash S. Immature gastric teratoma in a newborn: a case report. J Neonatal Surg, 2016, 5(2):21. PMID: 27123405 PMCID: PMC4841377

[15] Dunlap JP, James CA, Maxson RT, Bell JM, Wagner CW. Gastric teratoma with intramural extension. Pediatr Radiol, 1995, 25(5):383–384. https://doi.org/10.1007/BF02021715 PMID: 7567273

[16] Bowen B, Ros PR, McCarthy MJ, Olmsted WW, Hjermstad BM. Gastrointestinal teratomas: CT and US appearance with pathologic correlation. Radiology, 1987, 162(2):431–433. https://doi.org/10.1148/radiology.162.2.3541031 PMID: 3541031

[17] Beavers AJ, Khan A, Uddin N, Weakley DL, Setoodeh S, Pfeifer CM. Multimodal depiction of a rare immature gastric teratoma from fetus to infant. Radiol Case Rep, 2018, 14(3):372–376. https://doi.org/10.1016/j.radcr.2018.12.003 PMID: 30581526 PMCID: PMC6302251

[18] Bourke CJ, Mackay AJ, Payton D. Malignant gastric teratoma: case report. Pediatr Surg Int, 1997, 12(2–3):192–193. https://doi.org/10.1007/BF01349998 PMID: 9156857

[19] Heifetz SA, Cushing B, Giller R, Shuster JJ, Stolar CJ, Vinocur CD, Hawkins EP. Immature teratomas in children: pathologic considerations: a report from the combined Pediatric Oncology Group/Children's Cancer Group. Am J Surg Pathol, 1998, 22(9):1115–1124. https://doi.org/10.1097/00000478-199809000-00011 PMID: 9737245

[20] Ukiyama E, Endo M, Yoshida F, Tezuka T, Kudo K, Sato S, Akatsuka S, Hata J. Recurrent yolk sac tumor following resection of a neonatal immature gastric teratoma. Pediatr Surg Int, 2005, 21(7):585–588. https://doi.org/10.1007/s00383-005-1404-y PMID: 15928937

[21] Wagener C, Menzel B, Breuer H, Weissbach L, Tschubel K, Henkel K, Gedigk P. Immunohistochemical localisation of alpha-fetoprotein (AFP) in germ cell tumours: evidence for AFP production by tissues different from endodermal sinus tumour. Oncology, 1981, 38(4):236–239. https://doi.org/10.1159/000225557 PMID: 6165946

[22] Junhasavasdikul T, Ruangwattanapaisarn N, Molagool S, Lertudomphonwanit C, Sirachainan N, Larbcharoensub N. Immature gastric teratoma in an infant: a case report and review of the literatures. Clin Case Rep, 2016, 4(10):962–967. https://doi.org/10.1002/ccr3.654 PMID: 27761247 PMCID: PMC5054471

[23] Yamashita N, Kanai H, Kamiya K, Yamada K, Togari H, Nakamura T. Immature teratoma producing alpha-fetoprotein without components of yolk sac tumor in the pineal region. Childs Nerv Syst, 1997, 13(4):225–228. https://doi.org/10.1007/s003810050072 PMID: 9202859

[24] Marina NM, Cushing B, Giller R, Cohen L, Lauer SJ, Ablin A, Weetman R, Cullen J, Rogers P, Vinocur C, Stolar C, Rescorla F, Hawkins E, Heifetz S, Rao PV, Krailo M, Castleberry RP. Complete surgical excision is effective treatment for children with immature teratomas with or without malignant elements: a Pediatric Oncology Group/Children's Cancer Group Intergroup Study. J Clin Oncol, 1999, 17(7):2137–2143. https://doi.org/10.1200/JCO.1999.17.7.2137 PMID: 10561269

[25] Gupta DK, Srinivas M, Dave S, Agarwala S, Bajpai M, Mitra DK. Gastric teratoma in children. Pediatr Surg Int, 2000, 16(5–6):329–332. https://doi.org/10.1007/s003830000390 PMID: 10955556

Corresponding author

Min Jung Jung, MD, Department of Pathology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, 170 Jomaru-ro, Wonmi-gu, Bucheon-si, 14584 Gyeonggi-do, Korea; Phone +82–32–621–5962, Fax +82–32–621–5961, e-mail: mj@schmc.ac.kr

Received: January 6, 2020 *Accepted: April 21, 2020*